interpreted as a developmental error caused by the abnormal proliferation and behaviour of embryonal cells.

R. M. S.

SENSORIMOTOR NEUROLOGY


Congenital stenosis of the aorta is not a very uncommon anomaly. Circulatory disturbances with brain involvement incident to this malformation may occur. Autopsy of the patient under discussion verified this fact.

The patient presented not only a congenital stenosis of the aorta, but also a bicuspid aortic valve. The latter is a common finding and a frequent site for the superimposition of an endocarditic infection. The frequent involvement of the aortic flaps of the mitral valves and the secondary bacterial invasion of such congenital anomaly of the heart are well illustrated in this case. It is to be noted that one of the terminal causes of death in this case was a subacute vegetative endocarditis.

The mental symptoms, the hemiplegic attacks and death of this patient occurred between the second and third decade of life. Clinically he presented cardiac signs of a congenital anomaly of the heart together with excessive pulsations in the vessels of the neck, the suprasternal notch and palpable pulsations in several intercostal spaces. There was a marked difference in the blood pressures of the lower and upper extremities together with definite signs of cerebral involvement. He conformed categorically to the 'textbook' description of cases of coarctation of the aorta. There were evidences of arteriosclerosis in spite of the patient's youth and a bilateral simple optic atrophy. There was no evidence of either congenital or acquired syphilis.

Aphasia, dementia, focal and bilateral paralysis resulting from previous attacks of hemiplegia, incontinence, athetosis and muscular contractions were some of the manifest neurological signs the patient presented. When one considers the various sensory and other motor disturbances which may have been present, but which could not be elicited owing to the mental state of the patient, it is at once apparent that almost any and all neurological complications may be encountered in a case of this kind.

The neurological signs and symptoms are not primarily neurogenic in origin, but are secondary to the vascular complications such as cerebral haemorrhage, infarct, embolism, aneurysmal rupture, encephalomalacia and cerebral arteriosclerosis. The greater part of the anatomical diagnosis in this case is concordant with such a contention.

Although congenital cardiovascular anomalies may or may not be associated with congenital degenerative diseases of the nervous or other systems, the characteristic findings of a stenotic aorta previously mentioned, even in the presence of a heterogeneous conglomeration of neurological signs
and symptoms, should aid one in ascertaining the true nature of the clinical picture presented.

R. G. G.

[86] Calvarial hyperostosis and the accompanying symptom-complex.—


Moore recognizes four types of thickening of the calvaria; hyperostosis frontalis interna, nebula frontalis, hyperostosis calvariae diffusa and hyperostosis frontoparietalis.

In the first type the overgrowth of cancellous bone lies on the inner table and is covered on its intracranial aspect by a smooth lamella of compact bone. There is no evidence of an inflammatory process. This deposit of bone is increased in density, as seen in roentgenograms. Increase in density progresses from the inner table outwardly through the diploe and the hyperostotic deposit is progressive; roentgenographically it can be divided into degrees of development. Morphologically, it may be either nodular or sessile. In certain cases the orbital plate of the frontal bone is chiefly, if not exclusively, involved, and this change may be combined with calcification in the falx cerebri.

The second type of calvarial change consists of a triangular or ellipsoid area of increased density located in the squama frontalis, extending downward from a base on the sagittal plane. There are no projections from its intracranial surface, and it is less clearly defined than the preceding type of hyperostosis. It also has a progressive development.

The third type of hyperostosis is one in which there is a generalized increase in thickness of the entire vault, with an increase of density of the diploe which is in excess of that to be anticipated from mere increased volume of bone.

The fourth type, which is perhaps but a step in the development of the third, has the maximum thickening of the diploe at approximately the centre point of the parietal and squamous portions of the frontal bones. This type of hyperostosis produces a gentle grooving of the vault at the site of the sagittal and coronal sutures.

In each of the four types there is bilateral symmetry, both in extent and degree, of the osseous changes. There is no change in the outer table of the skull, which remains regular and smooth. The skull does not increase in size, so that the increased volume of the bone has to be accommodated at the expense of the capacity of the cranial cavity. The incidence of all types was 3-5 per cent.

The symptoms include headache, often referred to the forehead, obesity of the rhizomelic type, muscular weakness, epileptiform seizures, mental dullness and disturbance of gait. The symptom-complex and the roentgen evidence are so characteristic that, when one is observed, the appearance of
the other may be foretold in a substantial proportion of cases. Probably the symptoms antedate the osseous changes.

The differential diagnosis of the disorder has to be made first from osseous dystrophies. The unequal incidence in the two sexes (98 per cent. in women) separates these conditions from other osseous dystrophies, practically all of which occur with approximately the same frequency in the two sexes. The condition occurs rather late in adult life and may possibly be a metabolic disease in which, as far as is known at present, only fat and calcium metabolism is at fault.

R. M. S.


Carr's series of 17 cases of hyperostosis of the frontal bones all occurred in women, and in more than half obesity was a prominent feature. Increased sugar tolerance, low basal metabolic rates, menstrual disturbance and headache were also noted.

In several cases striking improvement followed aminoacid therapy, gelatin being used for this purpose.

To assist in demonstrating the clinical features, Carr relates the histories of three cases claimed to be typical of the syndrome; the information given is, however, too meagre to be of much value.

R. M. S.


A report of a family is presented in which five members suffered from convulsive seizures or migrainoid attacks with focal manifestations. Roentgenograms of the skull in two cases disclosed multiple areas of intracranial calcification. Pathological examination of the lesions in one case showed them to be telangiectases of the brain.

The clinical diagnosis of cerebral telangiectases is made with difficulty. The presenting symptom is usually epilepsy. Focal attacks and paralysis are frequent. The long duration of symptoms and the absence of intracranial hypertension will exclude neoplasm. Telangiectases may be suspected from the family history, the existence of extraneural vascular malformations, or possibly the presence of peculiar areas of intracerebral calcification seen in the roentgenogram. Attention is called to a report by Fritzche of a familial disease of the nervous system characterized by diffuse calcification in the grey matter of the brain—cerebral cortex, basal nuclei and dentate nucleus.
In their review of cases of multiple telangiectases of the nervous system the authors omit all reference to the cases recently described in this country which are characterized by amnesia, nevus in the territory of the fifth cranial nerve and contralateral infantile cerebral hemiplegia.

R. M. S.


The irregularity of the anatomico-pathological findings in a few cases of prefrontal tumours occurring with the akinetic-hypertonic syndrome caused the author to study systematically from a clinical aspect the anatomy and pathology of a case of his own.

The results of these investigations have induced the author to put forward an interpretation which regards the intervention of the frontal lobe in the production of this syndrome in the light of a pathological modification of descending stimuli, achieved in the first place by a simple dysfunction of subcortical structures and later by a structural alteration.

R. G. G.

[90] On tumours of the septum pellucidum (Sui tumori del setto pellucido).—F. Cardona. Ric. di pat. nerv. e ment., 1936, 47, 265.

These tumours are very rare, but two cases of glioma are described. They showed characteristically confusion and epileptic convulsions and in one case subsequent depression. In the second case there was a post-convulsive hemiparesis on the left and a left abducens paralysis. This tumour touched the internal capsule, hence the hemiparesis, the abducens paralysis being explained by increased intracranial pressure.

Tumours in this situation may be unrecognized, since the psychic disturbance is hardly diagnostic. Localizing symptoms are only fortuitous and due to chance pressure on neighbouring structures.

R. G. G.


The author describes three cases presenting the classical signs of intracranial pressure. In addition there were signs usually associated with cerebellar lesions and defects in the lateral parts of the visual fields.

The author refers to Cushing’s work, which shows that, in view of the
paths of the optic radiations, defects of the visual fields, especially in the upper quadrantal areas, may occur in temporal lesions. He then goes on to discuss whether apparent cerebellar signs, especially if irregular and referred to both sides of the body, can be due to frontal or temporal lobe lesions.

He believes that the explanation in the case of the temporal lobes is that there may be a suprasegmental centre for the vestibular apparatus situated in the temporal lobe.

R. G. G.


The literature of the subject is reviewed and the diagnostic importance of this sign is assessed. A case is described in which various grasping phenomena were observed in the period before death.

It is necessary to try to define the nature and localization of the phenomena, which are still in some uncertainty.

R. G. G.


This article, describing experimental work and the clinical observation on a case, illustrates the importance of spatial perceptions in psychic ‘make-up,’ and claims that it is a formal constituent of the power of perception and that interference with spatial perceptions will influence both gnosis and praxia.

R. G. G.


This paper gives in rough outline a psychological analysis of motor behaviour in relation to the problem of automatism. An attempt is made to apply Jackson’s principle of ‘reduction to a more automatic condition’ in cerebral lesions to cases of apraxia of different kinds. A psychological analysis of constructional tests is given, and the relation of constructional apraxia to other forms of apraxia is illustrated by cases and discussed. The special test of ‘free construction,’ in contrast to constructing with a given pattern, is described and its psychological significance analysed.

C. S. R.
Following the removal of the left cerebral hemisphere for extensive infiltration by a tumour, clinical examination of the patient, a right-handed woman, yielded the following findings:

An elementary vocabulary was retained, which was partially increased by training in speech.

It was difficult to evaluate the mental capacity of the patient. She was more calm after operation, but was less willing to perform coordinated movements. She did experience emotional reactions, but they were not marked.

The functions of the cranial nerves were well preserved except for slight weakness of the right side of the face and the absence of the corneal reflex on the left.

The spastic paralysis on the right side was replaced by flaccid paralysis.

The presence of acute pain with motion of the joints or compression of the deep muscles demonstrated the existence of a centre of sensation below the cortex.

Studies of the vasomotor responses of the extremities by determinations of skin temperature did not show measurable alterations in peripheral temperature regulation on either the ipsilateral or the contralateral side.

R. M. S.

Three clinical cases are reported in deeply comatose patients, showing a particular type of reflex synergia.

Stimulation of the head and neck, of the upper extremities (with the exception of the thenar eminences and palms of the hands) and of the upper parts of the chest down to the borderline of D3 caused an extensor reflex of the upper extremities. Stimulation of all parts below D3 and of the palms and the thenar eminences produced flexor synergias. These reflex movements could be evoked only by nociceptive stimuli. The exceptional fact is emphasized that the reflex-responses varied according to the location of the stimuli in a regular manner.

The pathophysiological significance of this phenomenon cannot as yet be satisfactorily explained.

A survey of the literature indicates that reflex synergias of the upper extremities may exist already in lesions of the upper cervical cord without implying complete loss of voluntary innervation or consciousness. In such cases the reflex synergias have to be considered as mere spinal automatisms. The three reported observations, however, showed extensive destruction of
one or both cerebral hemispheres with evidence of increased intracranial pressure. The clinical picture resembled somewhat the phenomena seen in decerebrate animals; hence it is assumed that in addition to an undamaged spinal cord a fairly normal brainstem function is necessary to produce the types of reflexes described above.

R. G. G.


From the systematic radiological examination of the cranium of 25 cases of essential epilepsy the author found as follows:

In none of them was there any evidence of radiographic alteration from normal which could suggest any pathological endocranial lesion that might be responsible for the convulsive manifestations.

In five cases pathological alterations in the volume of the sella were noted (one showed a small sella, one too large a sella, two modifications of the posterior clonoid processes, and one epiphyseal calcification).

In eight cases the radiograms suggested intracranial hypertension. It is difficult, however, to be sure of the existence of hydrocephalus, and dilatation of vessels alone cannot be regarded as a definite sign.

Some cases show all these signs without any corresponding clinical manifestations.

R. G. G.


Several cases have been presented which correspond faithfully to the description of the syndrome designated as heaped small attacks or pyknolepsy.

From a review of the literature it seems that this type of seizure has a good prognosis and disappears at puberty, never to return.

It is probably not a disease entity with specific aetiology, but a type of reaction of the nervous system.

One can, therefore, only speak of a ‘pyknoleptic type’ of seizure, regardless of the aetiology.

Why such a reaction should have an apparent good prognosis is not clear.

R. G. G.


Three cases of vascular lesions in the hind-brain are described. They showed a contralateral hemianalgesia and hemithermanesthesia, including the face,
with intact sensory facial innervation, on the same side. Other signs pointed to the localization of the lesions at a point which interrupted the pain-temperature pathways behind the point of entry of the trigeminal nerve.

R. G. G.

[100] Hemiballismus on a luetic basis (Hemiballismus auf luischer Grundlage).

A man 45 years of age suffered from left-sided hemiballismus for only four weeks before death.

On microscopical examination definite changes were observed in the right corpus Luysii and in the corpus striatum. In many parts of the brain syphilitic endarteritis of the smaller vessels was also observed. The clinical symptoms had resulted from alterations in the cells of the corpus Luysii and corpus striatum. As a result of syphilis such changes had previously been observed only in cases of general paralysis. The patient showed no evidence of general paralysis but suffered from meningovascular syphilis of the nervous system, the infection having been acquired twenty-three years previously. Hemiballismus results not from stimulation but from ‘abolition of function’ (Ausfallssymptom) by destruction of cells in the corpus Luysii.

M.


A case of diffuse carcinomatosis of the dura is presented which simulates pachymeningitis hemorrhagica interna. Diffuse carcinomatosis of the meninges is a condition of some rarity. The literature is reviewed and eleven somewhat similar cases of this unusual manifestation of carcinoma are cited.

Diffuse carcinomatosis of the dura is secondary to a primary carcinoma elsewhere. A clinical diagnosis can occasionally be made by the findings of tumour cells in the cerebrospinal fluid. Severe headache with or without evidence of an intracranial metastatic lesion in persons with carcinoma or suspected malignancy should direct attention to the possibility of a diffuse or localizing metastatic carcinoma of brain coverings.

R. G. G.


The remarkable clinical fact in this case, namely, that the patient suffered little or no inconvenience from an extensive invasion of the substance of the
spinal cord by vascular hyperplasia, furnishes a striking example of the adaptability of the nervous structure to mechanical interference. The patient had complained from time to time of pain in the region of the shoulders and arms. With the exception of the pain which was regarded as 'rheumatic,' the patient stated that he was well until May 27, 1933, when he awakened with severe pain in the right arm, followed by weakness and complete loss of motion in the legs. He was admitted to hospital three days later, with complete motor and sensory loss in the affected limbs, with retention of urine, bloody spinal fluid under pressure of 8 mm. of mercury, and subsequently very severe pains in the legs, made worse by passive movement, later vomiting and respiratory difficulty. The malady terminated in death eight days after the onset of the paralysis, the result of secondary haemorrhage originating in an embryonal developmental anomaly—an angioplastic hypertrophy.

R. G. G.


In the two cases reported in this paper an affection in the spinal cord rapidly developed. The first showed retrobulbar neuritis and a special eye examination could not be made in the other as this patient died just after being admitted into hospital. In both cases a sharply defined focus was observed in the optic chiasma in which the nerve fibres were demyelinated. Probably such alterations may be observed in different affections of the nervous system, but some cases, as in the first case described in this paper, may belong to a special affection with characteristic signs.

M.


The author describes a case of a boy observed for eight years who from birth showed an unstable organization of both structure and function of the central nervous system and early developed a condition which from its slow course and limitation to signs of involvement of the pyramidal tract the author diagnosed as a primary spastic paralysis.

R. G. G.

A clinical description of the case is given. The course was slow and peripheral and cranial nerves seemed to be involved, and the vesical sphincters were affected. The cerebrospinal fluid showed increase in protein and globulin, and syphilitic reactions were negative. Histological studies showed alterations in the cells and fibres of the central nervous system, in the spinal ganglia and in the spinal roots and cranial nerves. The discovery of lesions in the spinal ganglia is a factor hitherto undescribed in amyotrophic lateral sclerosis. There was also degeneration of the optic nerve, which has not previously been described.

R. G. G.


Three sisters in a fairly large family suffered from the complaint. The muscular lesions eventually produced an almost complete immobility. The only lesions found were in the locomotor muscles, not in the vegetative muscles. There was no damage to the nervous system, central or peripheral, or to the endocrine or any other system.

The death of the patients took place rapidly in an acute delirium.

R. G. G.


A case of myotonia congenita and a case of dystrophia myotonica are described. The myotonia, which appeared to be the same in the two, was studied electromyographically.

The after-contraction of myotonic muscles, which persists after the cessation of voluntary effort or brief mechanical stimulation, is accompanied by action currents. This indicates that the phenomenon is neurogenic rather than myogenic.

The evidence presented favours the hypothesis that the after-contraction of myotonia is of reflex origin and is due to the persistent discharge of hyperexcitable sensory end-organs in the muscle.

Injections of ergotamine tartrate, pilocarpine hydrochloride and epinephrine hydrochloride have proved ineffective in relieving the myotonic
condition, although calcium gluconate and calcium chloride have significantly reduced the amount and duration of the after-contraction.

R. M. S.


From the study of 82 cases in which multiple neuritis was an outstanding clinical entity, and in many of which there were mental symptoms, it was found that digestive disturbances were constantly present. A logical explanation for the lack of intake, failure of proper digestion or assimilation could be found in nearly every case. The varieties of clinical syndromes closely approximated those seen in such deficiency diseases as beriberi, pellagra, and sprue, which are accepted as being due to a lack of one of the vitamin groups. The known pathology of the nervous system in these syndromes compares favourably with that seen in the accepted deficiency diseases. Finally, there was the therapeutic test of improvement when the patients were placed on a high vitamin diet.

From the above observations the author believes that the failure to ingest the required amount of vitamins, or the inability to digest or assimilate the vitamins ingested, is responsible for the neurological syndromes presented by the patients.

He can offer no definite evidence as to any specific vitamin that is responsible for these changes in the central nervous system. Since the syndromes offer both clinical and pathological fragments of deficiency diseases which are accepted as being due to different vitamins, he supposes that in many instances the lack of more than one type of vitamin is responsible for the pathology. Until this subject has been more thoroughly investigated, these patients should be treated by a full diet rich in vitamins, especially B₁ and B₂.

R. G. G.


Undulant fever, characteristically a generalized infection, may show evidence of localization of the disease process. Uncomplicated undulant fever is almost always accompanied by some nervous system symptoms, apparently due to the action of a bacterial toxin which may have a special affinity for the central and peripheral nervous systems. Occasionally there is an actual invasion of the nervous system by the causative organism, with a resulting encephalitis, meningitis, or myelitis. Focal localization of the
infection within the nervous system may result in aphasia, hemiplegia, hypertrophic pachymeningitis, muscular atrophies and pareses, or transverse myelitis. The diagnosis is suspected when characteristic neurological signs and symptoms are found in a patient known to have undulant fever, but examination of the cerebrospinal fluid is necessary to ascertain the diagnosis. The spinal fluid characteristically shows increased pressure, pleocytosis, increased albumin, and decreased glucose and chlorides, but for confirmation of the diagnosis the organism of undulant fever must be isolated from the fluid. Pathologically, there is a subacute, non-specific meningitis, which may not be localized, and a diffuse or localized myelitis or encephalitis, characterized by perivascular infiltrations and ganglion cell degeneration. The nervous system complications may be the first evidence of systemic undulant fever or may not occur until months after the onset of the infection. The reported cases have all occurred in young individuals. Treatment is limited to general measures and various types of sera, the efficacy of which has not been definitely determined. Repeated lumbar punctures may be of value. These complications are rare, serious, but not necessarily fatal. Their prognosis, to a certain extent at least, may be related to the relative pathogenicity for man of the variety of Brucella melitensis involved.

A case of undulant fever meningoencephalitis has been presented. The clinical picture was characterized by headache, vertigo, diplopia, tinnitus, transient paralyses, and nuchal rigidity. The diagnosis was confirmed by the spinal fluid findings, and by the isolation of Brucella melitensis, var. abortus, from the fluid. Some improvement followed treatment by means of palliative measures and antiserum.

R. G. G.

[110] Contribution to the study of acromicria (Contributo allo studio dell’acromicria).—M. Bergoni. Riv. di pat. nerv. e ment., 1936, 47, 125.

Acromicria is the antithesis of acromegaly and is characterized by a smallness of the extremities and a tendency to fragility of the bones. There is an alteration in the quality and diminution of quantity of the eosinophils in the anterior lobe of the pituitary. There is a large quantity of colloid in the intermedia of the pituitary and a basophil infiltration of the pars nervosa.

The thyroid is small and there is a great scarcity of colloid in this gland. There is in fact a definite hypothyroidism.

R. G. G.


In 400 cases of neurosyphilis, the males showed 38 per cent. asymptomatic, 56 per cent. parenchymatous, and 6 per cent. meningovascular neuro-
Chymatous, and syphilis; nulliparae showed 63 per cent. asymptomatic, 29 per cent. parenchymatous, and 8 per cent. meningovascular neurosyphilis; multiparae showed 54 per cent. asymptomatic, 38 per cent. parenchymatous, and 8 per cent. meningovascular neurosyphilis. When arranged in the age groupings the nulliparae showed a preponderance of cases in the third decade (ages 20 to 29) and a very small percentage after 40 years of age. In contrast, the males and multiparae showed the peak in the fourth decade (ages 30 to 39) with a much higher incidence after this period than in the case of the nulliparae. A detailed analysis of the incidence of each clinical type in each age grouping is shown in tables.

R. G. G.

**PROGNOSIS AND TREATMENT**

[112] *Epilepsy and surgical therapy.*—WILDER PENFIELD. *Arch. of Neurol. and Psychiat.*, 1936, 36, 449.

Surgical therapy demands the most exhaustive preliminary study of the pathological anatomy involved, and no surgical procedure should be countenanced unless it is directed by rational analysis of the individual aetiological problem. In idiopathic epilepsy each seizure is probably initiated by a discharge in the grey matter, which is just as focal as that in the cases of epileptiform seizures which result from a gross lesion of the brain. There may be nothing in the outward manifestation to distinguish one form of convulsion from the other. But the initial clinical problem is to discover whether or not a gross lesion exists in the brain as well as what pathological influences may play on it. When history, examination or pattern of attack suggests the possibility of a focal lesion, an encephalogram should be made and an attack observed.

In essential epilepsy there is no gross organic defect of the brain, but there is nevertheless a common organic abnormality, which may be described as cerebral vasalability. This irritability of cerebral vessels is found also, although to a less extent, in focal epilepsy with a gross lesion of the brain.

Conclusions regarding the more important methods of surgical treatment proposed for essential epilepsy may be summarized as follows:

1. Cervicothoracic sympathetic ganglionectomy has failed, except perhaps in the occasional case in which the condition is associated with obvious abnormality of the sympathetic nervous system.

2. Removal of the carotid body and denervation of the carotid sinus are as yet without theoretical justification except in the rare case of demonstrably abnormal carotid sinus reflex. Nevertheless, the practical results secured by Lauwers demand further consideration.

3. Subtemporal decompression should be carried out only occasionally as an incident to craniotomy undertaken for other purposes or in the rare
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