CORNELL UNIVERSITY
MEDICAL COLLEGE
NEW YORK CITY

JUL 30 1940

THE
JOURNAL OF MENTAL SCIENCE

[*Published by Authority of the Royal Medico-Psychological Association.*]

No. 363 [NEW SERIES No. 327.] JULY, 1940. VOL. LXXXVI

Part I.—Original Articles.

UNILATERAL HYDROCEPHALUS.

REPORT OF TWO CASES OF THE NON-OBSTRUCTIVE TYPE.

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(Received March 16, 1940.)

A SINGLE small aperture connecting each lateral ventricle with the dorso-cephalic portion of the third ventricle provides the sole means of escape for cerebro-spinal fluid, and consequently it follows that any interference with its patency will seriously retard the outward flow of ventricular fluid. Complete, or even partial, closure of the foramen leads to a rapid increase in the volume and pressure of imprisoned ventricular fluid so that a condition of internal hydrocephalus is soon established. Usually both foramina are occluded, the hydrocephalus being therefore bilateral and the ventricles more or less symmetrically enlarged. In rare instances, however, only one foramen is obstructed, in which case the dilatation will, of course, be confined to the ipsilateral ventricle. Unilateral hydrocephalus of this obstructive type is commonly caused by inflammatory conditions in the neighbourhood of the foramen, or by pedunculated tumours attached to the choroid plexus which enjoy a degree of movement sufficient to permit intermittent or permanent blockage of the foramen of Monro. It is, however, possible to find examples of unilateral hydrocephalus in which the outflow of cerebro-spinal fluid through the foramen of Monro is unimpeded, and in these the cause of the ventricular dilatation is to be sought in some diseased condition of the cerebral wall which has become so weakened as to be unable to resist even normal ventricular pressure. Such expansion may be either limited to one part of the lateral ventricle, or general, involving the entire body with its horns.

https://doi.org/10.1192/bjp.86.363.591 Published online by Cambridge University Press

In illustration of this second non-obstructive type of unilateral hydrocephalus the following two cases may be quoted :

CASE 1.—When admitted to hospital on 2.ix.14, A. W—, a female, aged 14, was noted to have the physical development of a child of five ; unable to articulate or to occupy herself in any way, dirty in habits and subject to severe epileptic fits, she was a typical example of profound idiocy. In 1924, when transferred to Leavesden Hospital, she was found to have spastic paralysis of right upper and both lower limbs. Corneal clouding on both sides and traces of notches on the upper central incisors suggested the presence of congenital syphilis, and after a course of novarsenobillon her fits became less frequent, but otherwise no improvement was noted. Ten years later the paralysis had become more marked, especially on the right side ; the pupils reacted normally, the deep reflexes were exaggerated and the plantars could not be obtained. The Wassermann reaction was negative in the blood and cerebro-spinal fluid ; the pressure of the latter was normal. On 21.xi.34 the patient had a succession of fits, the movements being almost confined to the left side. Death occurred at 7.20 p.m. on 24.xi.34.

An autopsy was performed on 26.xi.34 at 10 a.m. The body was that of an undersized adult female. The limbs on the right side were smaller than on the left, the right arm showing contracture at the elbow-joint, and the right leg a marked degree of talipes equino-varus. The calvarium was increased in thickness and density, but showed no asymmetry. The dura was normal. The brain was microcephalic, weighing 870 gm. The pia-arachnoid showed intense congestion over the frontal and parietal gyri of the right cerebral hemisphere, but without thickening or opacity. On the opposite side it showed neither congestion nor thickening, having, indeed, an almost normal appearance.

On the right cerebral hemisphere the convolutions were simple, but showed no atrophy. Those on the vertex of the left hemisphere were of a yellowish-brown colour, flattened to a marked degree and pressed tightly together (Figs. 1, 2). Their staining and flattening into ribbon-like areas was particularly marked in the territory supplied by the middle cerebral artery. On palpation the convolutions had an almost leather-like consistence. Division of the attenuated corpus callosum allowed the escape of a large quantity of clear cerebro-spinal fluid and when the two hemispheres were separated the left lateral ventricle was seen to be enormously dilated and to have lost all semblance of its normal shape (Fig. 3). As is usual in hydrocephalus, the white matter had yielded more readily than the grey, being hardly recognizable, and thinned to a mere shell which in some areas was actually translucent. In the neighbourhood of the frontal, temporal and occipital poles the grey and white matter were of an almost normal thickness, the convolutions in these localities being relatively well formed. The ependymal lining of the ventricles was for the most part smooth and shiny, but here and there it presented a wrinkled appearance ; the choroid plexus was thickened and bound down. The right lateral ventricle showed a very slight degree of dilatation, but with a perfectly normal ventricular lining. The third and fourth ventricles were normal and the Sylvian aqueduct connecting them showed no dilatation. No evidence of recent or old inflammation could be found at the base of the brain and in this situation the arteries showed no evidence of disease. The cerebellum showed marked asymmetry, its right hemisphere being notably smaller than the left. In the medulla the right pyramid and the left inferior olive were reduced in size.

Microscopic appearances.—Sections of cerebral cortex were made from the frontal and parietal regions of both cerebral hemispheres and stained for nerve cells, glia and myelin sheaths.

In the right cerebral hemisphere little abnormality could be found. There was no obvious disturbance of the cyto-architectonics, but in all laminae of the pre- and post-central gyri the nerve cells were poorly developed and fewer than normal (Fig. 4*a*). Apart from some increase in glial nuclei, no evidence of gliosis could be found.

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Over the left cerebral hemisphere the pia mater showed slight thickening; it contained numerous fibroblasts but no inflammatory cells and its vessels were normal.

In the left pre- and post-central gyri cortical devastation was almost complete, the total depth of the grey matter being about one-quarter of the thickness of the corresponding gyri on the opposite side (Fig. 4*b*). No nerve cells could be found

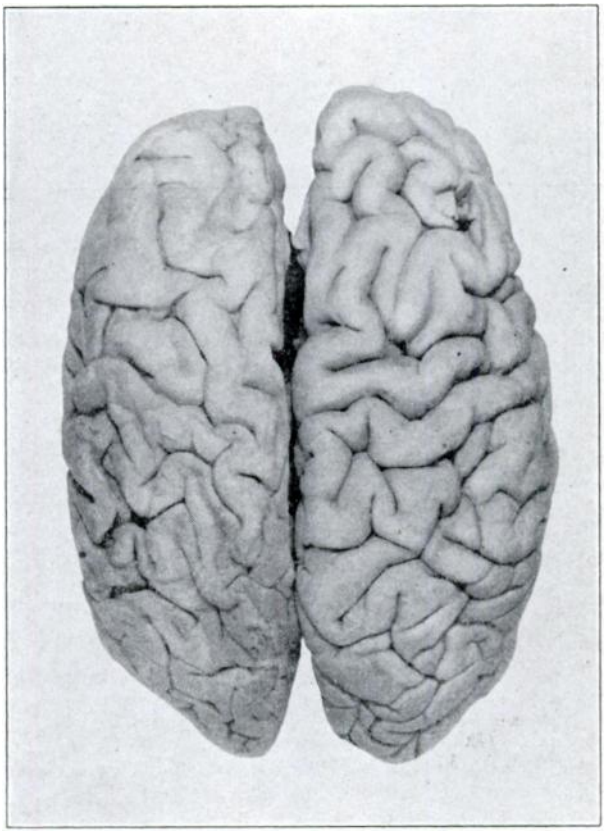


FIG. 1.—Cerebrum. The left hemisphere is smaller than the right and its convolutions markedly flattened. ⅔ actual size.

except in the deeper layers and here those which survived were immature, many of them resembling neuroblasts. Rod cells were numerous.

In the molecular layer numerous yellow pigment granules could be seen lying scattered between a horizontal row of dark staining neuroglial nuclei.

The white matter was likewise much reduced in thickness and contained a great excess of glial nuclei.

Sections stained by Holzer's method revealed an intense degree of gliosis (Figs. 4*c*, 5*a*). Immediately beneath the pia a thin superficial stratum of fine interlacing fibres could be seen merging into dense strands and sheaves at a deeper level.

In the white matter the degree of gliosis, stronger and denser than in the grey matter, was most striking. Gliosis was also dense in the ependymal lining of the

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ventricle. The blood vessels in the grey matter appeared to be fewer than normal, but showed no structural abnormality.

In certain areas other changes were found. Thus, in the convolutions lying anterior to the precentral gyrus many areas could be found in which a loss of all components of the nerve parenchyma in the second and third cortical layers was accompanied by an imperfect replacement by fibrous glia and the establishment of numerous holes of varying size; the appearances were, in fact, typical of status spongiosus (Fig. 5*b*).

In the more normal convolutions of the temporal and occipital poles it was possible to find areas in which nerve-cell lamination was retained, but even here a marked degree of gliosis was present.

Cerebellum.—The two unequal lateral lobes showed no obvious changes. For the most part the molecular and granular layers were well developed and the Purkinjë cells had suffered no numerical loss, but here and there areas existed (Fig. 6) in which all three layers had undergone a striking degree of atrophy, while contiguous folia were completely preserved.

Medulla oblongata.—The right pyramid tract showed a marked degree of sclerosis, no myelin sheaths being found in Weigert-Pal preparations. The left inferior olive was more slender and contained fewer nerve cells than the right.

The blood vessels of the medulla and ependyma lining the fourth ventricle were normal.

Summary of microscopic appearances.—The convolutions of the right cerebral hemisphere showed a marked poverty of nerve cells, but no disturbance of lamination and no gliosis. Those of the left cerebral hemisphere showed an almost complete destruction of nerve cells, associated with a replacement fibrillary gliosis and the establishment of status spongiosus.

CASE 2.—The second case was that of a male idiot aged 19, who, from an early age had shown weakness of the right arm. In addition he displayed the clinical features of osteochondro-dystrophy, or, as it is now called, dysostosis multiplex. The pathological findings of this case have been fully reported elsewhere (Ashby, Stewart and Watkin, 1937), and it is only necessary here to refer to the findings in the cerebrum.

The brain was of normal consistence and after fixation weighed 1,043 gm. The cerebral hemispheres were asymmetrical, the left being broader and flatter and 102 gm. lighter than the right. Occupying the stem of the Sylvian fissure and the gyri surrounding it and covered by thickened and opaque pia-arachnoid a large area of cortical atrophy could be seen.

The right cerebral hemisphere showed two small areas of thickened pia-arachnoid, but without cortical atrophy, one occupying the horizontal ramus of the Sylvian fissure and the other in the prefrontal region. On division of the two hemispheres a condition of unilateral hydrocephalus was revealed, the left lateral ventricle showing a very marked degree of dilatation affecting equally the body and its horns (Fig. 7). Save for a small area overlying the caudate nucleus there was no evidence of ependymitis. Attached to the choroid plexus in the body of the ventricle was a small firm rounded tumour about the size of a hazel nut, and from its position it was obvious that it could not have blocked the foramen of Monro. Another tumour, occupying the same position and of the same size, was present on the choroid plexus of the otherwise normal right lateral ventricle.

A vertical coronal section through the left cerebral hemisphere in the plane of the middle commissure showed that the area of softening was sharply confined to the cerebral cortex, with, however, marked thinness of the underlying white matter. On this side, too, the basal ganglia were much smaller than in the opposite hemisphere.

The cerebellum was symmetrical with a normal fourth ventricle. Microscopic examination of the area of cortical atrophy showed a complete absence of cortical cells and a marked excess of fibrous glia. Elsewhere the changes, which bore a close resemblance to those of amaurotic idiocy, were similar in both hemispheres.

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Review of Cases Previously Published.

The earliest reference to unilateral hydrocephalus is to be found in a communication by Von Mohr (1842). In his first case, a mentally defective girl who had acquired a right-sided hemiparesis in infancy, there was a history of epilepsy. Death occurred in her twenty-first year. Examination of the brain revealed an atrophied left cerebral hemisphere with an enlarged lateral ventricle; that of the right hemisphere was slightly dilated. The wall of the left lateral ventricle was very thin and its ependymal lining thickened. The foramen of Monro was unobstructed.

His second case was that of an insane woman who died in her sixty-first year. For many years she had had a right-sided hemiparesis. At the autopsy the left lateral ventricle was found to be much enlarged, its wall thin and compressed. There was also cystic degeneration of the posterior portion of the left corpus striatum with enlargement of the descending ventricular horn. The right lateral ventricle was normal and there was no obstruction of either foramen of Monro.

White (1901) recorded the case of a woman, aged 74, with a history of recurrent attacks of hemiplegia of short duration. On the left side an internal unilateral hydrocephalus was found, with marked poverty of cortical nerve cells, glial overgrowth and other degenerative changes. The ventricular wall was thickened and the choroid plexus enlarged with a firm mass in its substance containing numerous hyaloid bodies. The foramen of Monro was unobstructed.

In another case of unilateral hydrocephalus reported by Spiller (1902) the patient, a female idiot, showed a left-sided paresis. Death occurred at the age of fourteen. The left cerebral hemisphere was normal. On the right side there was marked ventricular dilatation, the walls being extremely thin, so that the whole hemisphere was a mere sac. The dilatation appeared to be due to inflammatory changes which had partially closed the right foramen of Monro.

Two cases of partial dilatation of one ventricle were reported by Cramer (1905). In the first, an idiot with right-sided hemiplegia had been epileptic since his seventh year; death occurred at the age of twenty. At the autopsy a marked chronic leptomeningitis was found which was most noticeable over the left frontal and temporal lobes. The left anterior horn was very large and had expanded the wall of the hemisphere into a thin shell. The author assumed that in this case a meningo-encephalitis had weakened the cerebral substance, thus permitting dilatation of the horn.

The second case was that of a man, aged 23, who had been ill for three months with symptoms of increased intracranial pressure. There was a marked right hemiparesis and some weakness on the left side. Decompression afforded no relief. At the autopsy the right temporal lobe and the descending horn were

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found to be much enlarged. The dilatation was attributed to the presence of an inflammatory mass which had completely closed the entrance to the temporal horn. Though tubercle bacilli were not found, a tuberculous origin was suspected.

Weber (1906) in a study of hydrocephalus in general refers to five cases, none of which was entirely unilateral. In the first, a chronic diffuse leptomeningitis with cystic degeneration of the hemisphere wall had resulted in atrophy of the brain-tissue over the anterior horn.

In the second case a subcortical encephalitis, probably syphilitic in origin, had softened the wall of one lateral ventricle.

The third case showed encephalitis and softening in the neighbourhood of the dilated ventricle.

The fourth case showed a chronic diffuse leptomeningitis with adventitial overgrowth and sclerotic areas, especially over the left lateral ventricle. This was probably a case of congenital syphilis.

In the fifth case there was also chronic leptomeningitis with areas of gliosis and softening external to the dilated ventricle.

In each of these five cases the foramen of Monro was unobstructed. Weber believes that two factors operate in the production of this type of hydrocephalus —diminished absorption of cerebro-spinal fluid owing to the presence of a chronic leptomeningitis and disease of the hemisphere wall which permits the ventricle to enlarge.

Hunt (1916) records the case of a woman, aged 78, who had a left-sided weakness of 25 years' duration. The report is fragmentary, no information being given as to the state of the foramen of Monro.

Dandy (1919) reported a case of the obstructive type in which a pedunculated tumour was suspended in the left ventricle and periodically closed the foramen of Monro.

Dott (1927) describes a case of unilateral hydrocephalus which appears to be the first of its kind. A progressive unilateral hydrocephalus was found to be due to obliteration of the left foramen of Monro by scar-tissue. In this case an active infective process had become localized and shut off in the ventricular system. Cure was effected by excision of a window in the septum lucidum, the normal right foramen of Monro thus providing an exit for the cerebro-spinal fluid from both lateral ventricles.

Hinrichsmeyer (1922) records a case of local atrophy of the brain substance with a corresponding enlargement of one lateral ventricle. The patient was a feeble-minded boy with epilepsy and a left-sided paresis dating from birth. An operation performed in his tenth year revealed a hydrocephalic enlargement of the right ventricle. Marked thinning of the brain substance was found in the frontal region, the island of Reil being completely destroyed. The foramen of Monro was normal on both sides.

Much more informative is the paper by Winkelman and Eckel (1924).

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Their patient was a man, aged 62, with a history of intense alcoholism and a paralysis of four years' duration.

On admission he was found to have a right hemiplegia, motor aphasia and definite evidence of senile deterioration. He improved, but after an interval of two years was readmitted in a state of semi-stupor and with even greater spasticity than before. Death was caused by another paralytic "stroke." The more important autopsy findings were: generalized arteriosclerosis, interstitial myocarditis and chronic nephritis. The brain was soft with fluctuation and recent areas of thrombotic softening in the left parieto-occipital region, flattening of the convolutions and a generalized shrinkage of the left hemisphere. The pia-arachnoid was slightly thickened and the vessels were sclerotic. On sectioning the brain after hardening the right ventricular system was found to be dilated but not more than usually found in a senile brain. On the left side the posterior horn was markedly dilated and the cortex surrounding it thinned out. No actual obstruction was found, although the foramen of Monro was slightly narrower than on the opposite side. The anterior portion of the left ventricular system and the descending horn were moderately dilated. On microscopic examination the characteristic findings of an arteriosclerotic brain were found. The cortex on the posterior portion of the left hemisphere was much narrowed and so altered as to be unrecognizable. In some areas there was complete replacement by a glial scar, in others the replacement was less complete. The condition of the pia-arachnoid varied; over a comparatively normal portion it showed a moderate fibrosis without cell infiltration, while over the posterior horn of the lateral ventricle it showed both inflammatory and proliferative changes. In some areas there was adherence of the cortex to the pia-arachnoid with proliferation of glial fibres in the form of sheaths. The vascular changes were those usually found in senile brains.

Under the title, "A Case of Cerebral Hemiatrophy or Unilateral Hydrocephalus," Wilson (1925) relates the case of a Chinese imbecile girl, aged 22, who was reported to have had frequent epileptic seizures and impulsive outbursts. The neurological signs were: microcephaly, right hemiplegia with wasting and contractures, right homonymous hemianopia and dysarthria.

Death was due to pulmonary tuberculosis. At the autopsy the skull, dura mater, right cerebral hemisphere, cerebellum, pons and medulla were normal; the right lateral ventricle was of normal size. The membranes of the left cerebral hemisphere were thickened, opaque and almost devoid of blood-vessels. They could be stripped with difficulty, disclosing the remains of the convolutions as "little dimples." It was impossible to make out any distinction between grey and white matter, the substance presenting a white, jelly-like appearance. The lateral ventricle was enormously distended; the ependyma was quite smooth. No mention is made of the condition of the foramen of Monro. The left cerebral hemisphere weighed $3\frac{3}{4}$ oz., the right, 17 oz.

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A somewhat similar case was described by Noronha (1925). A mentally defective Hindoo, aged 30, subject to periodical epileptic fits, followed by mental excitement, showed paralysis of the right half of the tongue, the face and upper limb. Speech was slow, slurring and interrupted. On post-mortem examination the dura mater was found to be thickened, and on incision an unusual quantity of cerebro-spinal fluid escaped. The right cerebral hemisphere was of normal consistence and the structures forming it well defined. The whole of the cortex of the left cerebral hemisphere formed the wall of a huge saccular cavity, filled with milky white fluid. Though not well marked the convolutions were easily distinguished, and when the contained fluid was allowed to escape, the hemisphere collapsed like a half-inflated football.

Diaz y Gomez (1933) has recorded a case of fibroma in the posterior cerebral fossa which, by occluding the foramen of Monro, caused internal hydrocephalus on the left side. Recovery after surgical intervention in unilateral hydrocephalus is reported by Tinel, de Martel and Guillaume (1932) in the case of a child aged 10, the dilatation resulting from an inflammatory process affecting the foramen of Monro.

Discussion.

From the pathological standpoint two distinct types of unilateral hydrocephalus may be recognized. In the first, which differs but little from the bilateral form, there is obstruction of the foramen of Monro; in the second type there is no obstruction, but disease of the hemisphere permits a passive ventricular dilatation on the affected side. Dott (1927) considers that the term "unilateral hydrocephalus" should be reserved for those cases in which one ventricle, or part of it, becomes dilated by reason of an obstruction affecting its cavity or its outlet, and he suggests that the term is a misnomer when applied to conditions in which one lateral ventricle has become passively enlarged by reason of local atrophic shrinkage of the adjacent brain-substance, inasmuch as the accumulation of fluid maintains normal pressure conditions within the cranium and is therefore actually beneficial.

He does not, however, suggest an alternative title for the non-obstructive type, and it is obvious that if the term is to be restricted to ventricular dilatation of obstructive origin a general revision of the nomenclature of all pathological states to which the prefix "hydro" is employed will be necessary. Thus, since there is little evidence to show that the cerebro-spinal fluid contained in a dilated central canal of the cord is under increased pressure, the term "hydromyelia" will have to be discarded.

It may be remarked, too, that it has yet to be proved that there is no increase in intraventricular pressure at the stage when the pathological process involving the cerebral wall is active. Meningitis and encephalitis are frequent causes of hydrocephalus, and in their acute phases some rise in cerebro-spinal

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fluid pressure is invariable, so that the presence of a normal ventricular pressure, perhaps years after the onset of the disease, does not necessarily rule out the possibility of increased pressure at the time when the inflammatory process was active. Moreover, that such a rise in unilateral hydrocephalus does occur is suggested by the not infrequent presence of a slight degree of ventricular dilatation on the side on which there exists no cerebral atrophy or other evidence of gross cerebral damage. This was present in the first case now reported. To the writer it seems unwise to limit the use of a term so long sanctioned by usage, especially if it be employed merely as a descriptive term, unfettered by implications regarding the mode of production of the hydrocephalus.

Concerning the cause or causes of the weakening of brain substance in this form of hydrocephalus it is difficult to speak with any degree of certainty. It is possible that either the whole or part of the brain may be weakened by an encephalitis occurring in utero, while similar processes of perhaps syphilitic or tubercular origin may develop in early life.

However produced, the general shrinkage of brain substance is probably never sudden but extends over a long period of time, so that the changes found at autopsy may represent the end-results of a process sustained many years previously. Consequently, the absence of all signs of inflammation does not necessarily mean that they may not have been present at one time. In some instances, as in Case 1, a clue to the duration of the hydrocephalus is afforded by the presence of a crossed atrophy of the cerebellum, for such a finding is a clear indication of a congenital or early post-natal origin. That the hydrocephalus in Case 1 was probably acquired in childhood and not in infancy is shown by the absence of cranial asymmetry, for its development in early life would almost certainly have been associated with yielding of the membranous bones on the affected side.

The presence in certain areas of status spongiosus affords belief for the view that destruction of all components of the nerve parenchyma must have been both rapid and intense, but the cause of this destruction remains obscure. Certain clinical features—and notably the corneal clouding and notching of the incisors—strongly suggested a syphilitic origin, yet neither in the nervous system nor in the visceral organs were there changes indicating such an infection. In particular, the meningo-vascular tissues, which in neurosyphilis so often bear the brunt of the damage, showed no residual signs of inflammation. It is, of course, possible for the nervous system to escape luetic involvement even when stigmata of congenital syphilis are obvious, and it may be assumed that this case was of such a character, for neurosyphilis being " an extensive disease of slight intensity," it would be difficult to understand how a syphilitic process which destroyed so large an area of one hemisphere could fail to implicate the other.

The second case is of interest in that the presence of a small tumour attached to the choroid plexus at first glance suggested hydrocephalus of the obstructive

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type, but the presence of another tumour occupying the same position and of the same size on the choroid plexus of the otherwise normal lateral ventricle on the right side indicated that the cause of the dilatation was to be sought elsewhere. As already described, the convex surface of the left cerebral hemisphere was occupied by a large area of cortical atrophy covered by thickened and opaque pia-arachnoid, but its extent seemed hardly sufficient to account for the uniform dilatation of the whole cavity, unless one assumes that the lessened resistance of the hemisphere wall extended much further than the area of cortical degeneration.

Summary.

Unilateral hydrocephalus may result from obstruction at the foramen of Monro or from disease of the hemisphere wall which permits a passive dilatation of the ventricle. The clinical and pathological features of two cases of the latter type are described.

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