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HYDROCEPHALIC IDIOCY.

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HYDROCEPHALUS is a not infrequent cause of idiocy. Producing, as it does, pressure upon the brain substance, with consequent impairment of function, either from the compression alone or from actual destruction by atrophy of the tissues, we note among the multiform results of hydrocephalus, enfeeblement of the mind, varying from simple feeble mindedness to complete idiocy, as well as paralysis, convulsions, blindness, and the like.

Bourneville distinguishes three classes of hydrocephalic idiocy, viz.:

1. Simple or common hydrocephalus.
2. Scapho-hydrocephalus.
3. Symptomatic hydrocephalus.

The first class he again divides into two groups: (1) Simple hydrocephalus without malformation of the brain, and (2) simple hydrocephalus complicated with

malformations or lesions, such as absence of the corpus callosum or of the cerebellum. This latter variety may include either the ventricles alone, or, at the same time, the arachnoid cavity, in which case it may be at times encysted in the arachnoid cavity.

To the writer this classification is unnecessarily complex. For instance, the second group is distinguished merely because the head has a scaphoid shape. But, since in simple hydrocephalus there may be several varieties of cranial conformation, as will be noted later, it scarcely seems justifiable to make of scaphocephalus a distinct type. The writer would, therefore, divide hydrocephalus into two classes only, viz.:

I. Primary hydrocephalus.

II. Secondary hydrocephalus.

To the first group belong the cases of ordinary hydrocephalus of unknown pathology. In the second class are comprised such cases of hydrocephalus as are secondary to meningo-encephalitis or encephalic tumors.

The condition we are more immediately concerned with is primary hydrocephalus, known variously as chronic hydrocephalus, hydrocephalus internus, and "water on the brain." It consists of an excessive accumulation of fluid in the ventricles of the brain, which expands the cavities to a greater or less degree, sometimes enormously, thus thinning the brain substance surrounding the fluid, and increasing the size of the head by separation and spreading out of the bones. The disorder is congenital or acquired. If acquired in childhood there is enormous development of the cranium. If acquired later in life the expansion of the skull may be slight or imperceptible. Where defect or malformation of the brain coexists the fluid may be found not only

in the ventricular cavities, but also exterior to the brain, constituting the condition of hydrocephalic anencephalus. Occasionally spina bifida coexists.

ÆTIOLOGY.—In all cases of hydrocephalus a neuropathic basis is found. Direct heredity is rare, though it is sometimes noted. Bourneville mentions a mother and two children affected with hydrocephalus. We discover commonly among the ascendants various neuroses or psychoses. Alcoholism in the parents is particularly frequent. In twenty-two cases reported by Bourneville, sixteen showed alcoholism in the antecedents, two absinthism, two no alcoholic history, and in two no history could be obtained. Insanity, imbecility, epilepsy, and migraine seem to be especially common conditions among ascendants. Disease of the mother during pregnancy is a not infrequent determining cause, such as instanced by Bourneville in a case having variola during the sixth month of pregnancy. Other conditions—for example, emotional excitement and accidents during pregnancy—seem to exert an influence on its origin. In some instances no determining cause will be ascertained.

PATHOLOGY AND MORBID ANATOMY.—The true pathogeny of primary hydrocephalus is unknown. It is generally explained as being due to a chronic intraventricular meningitis, a congestion of the ependyma. But in many of these cases nothing abnormal is observed about the ependyma save thickening. It is possible that a careful study of the manner of secretion of the cerebro-spinal fluid and of the relations existing between the ependyma and the external serous membrane of the brain may help to elucidate the origin of the disorder; for there is some reason for believing that a sort of current of fluid flows from the ventricles into

the exterior serous cavity through the foramen of Magendie, the foramina of Mierzejewsky, and two other foramina which have been described but are of uncertain existence. The ventricular walls secrete the cerebrospinal fluid and the exterior serous cavity absorbs it, according to this theory. Thus, then, there may be three processes by which primary hydrocephalus may be induced: hypersecretion in the ventricular spaces, occlusion of the foramina mentioned, and disorder of the absorbent apparatus. An interesting study of the subject along this line might be made.

When the fluid begins to increase in the ventricles, these become dilated, as a rule equally, occasionally unequally from obliteration of the foramen of Monro. The dilatation may be restricted to the lateral ventricles, or may include the third and fourth also. With the distention of the ventricles compression of the brain substance takes place, giving rise to functional impairment of various kinds and degrees. With increase of pressure, atrophy of the compressed parts occurs. The sæptum between the ventricles may disappear and the brain envelope become thin as paper, so that the hydrocephalus is like one enormous cyst filling the cranial cavity. The basal ganglia and brain stem become flattened. Examination of the cerebral envelope shows atrophy and degeneration of cells and fibres. The distention may go on until the cerebral tissues and the membranes vanish almost altogether. The amount of fluid has been known to reach six, eight, ten, twenty, and even twenty-seven pints. The following is an instance in point (a case from the Randall's Island Hospital for Idiots, the autopsy of which I reported at the New York Pathological Society—see Proceedings, 1894, page 94):

A female child, aged eighteen months; hydrocephalus, whether congenital or acquired unascertained. Circumference of head, 51.5 ctm.; antero-posterior diameter, 18 ctm.; greatest transverse diameter, 15 ctm.; naso-occipital arc, 32 ctm.; binauricular arc, 34 ctm.

Blindness and nystagmus; widely gaping fontanelles; spastic diplegia; occasional convulsions, and just before death opisthotonus. At the autopsy sixty-four ounces of reddish serum were first removed by tapping the anterior fontanelle. The skull and dura were exceedingly thin. The falx cerebri had disappeared. Cutting through the thin dura, nothing was to be seen in the great cavity of the head of any brain proper. The membranes usually covering the cerebrum had disappeared with that organ. At the base of the skull the floors of the ventricles and the basal ganglia stood out promi-

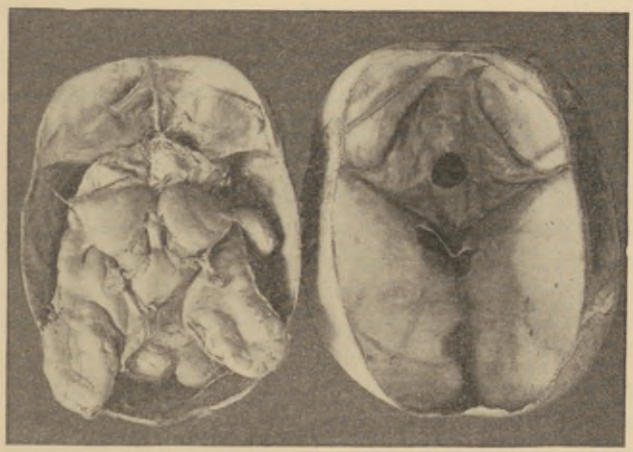

FIG. 1.—The cranial cavity in hydrocephalus, showing basal ganglia and cerebellum intact, but only vestiges of the cerebrum (parts of the temporo-occipital lobes).

nently, and back of these parts, lying on the tentorium, were the only vestiges of a cerebrum, parts of the two

occipital lobes. On removing the tentorium, the cerebellum was found to be of about normal size. Microscopical examination showed degeneration and atrophy of the lateral columns of the cord (Fig. 1).

In this case, then, we have to do with distention and atrophy of the encephalon pushed to its greatest extreme.

Case IV, in a series of autopsies by Bourneville, is a good illustration of the nature of the process of compression and atrophy: A girl, a complete idiot, died at the age of about two years. Five hundred grammes of fluid were found in the brain cavity, the brain envelope having become merely a sac of varying thickness. For instance, in the right hemisphere, over the whole of the temporo-occipital region, the wall of cerebral substances was but a millimetre in thickness, and at one place here near the fissure of Sylvius, the brain substance was absent altogether at a space of four centimetres in diameter, closed merely by a fine meningeal veil. In this case, then, the process of complete atrophy of the brain was arrested by death.

As the ventricular cavities dilate, pushing the brain envelope with them, the skull cavity is distended, and the cranial bones are separated, made thinner, and expanded in area. The enlargement of the head is directly proportional to the youth of the patient. Cases beginning before or shortly after birth will present greater expansion of the cranial cavity than such as have a later origin. Sometimes some sutures give way and others become synostosed. Where sutures are separated, Wormian bones often form, or a membranous connection is established between the cranial bones.

Occasionally in these cases of primary hydrocephalus, the defects of brain substance are not due to pressure-

atrophy, but there is an associated condition of malformation or defect. Thus, in an autopsy of Bourneville's on a girl, about thirteen years of age, with congenital hydrocephalus, idiocy, and epilepsy, the hemispheres of the cerebellum were totally absent, the cerebellum being represented by the vermis, which was the size of a pigeon's egg. Perhaps such a defect and others which have been described, are due to a pressure-atrophy beginning very early in fœtal life.

As regards the pathology of secondary hydrocephalus we possess more definite knowledge. In this the internal hydrocephalus is caused by obstruction of the veins of Galen or by obliteration of the foramina of Monro, Magendie, or Mierzejewski. Common causes are tumors of the cerebellum, such as sarcomata and tubercles. Meningitis may act in the same way. The amount of hydrocephalus, ventricular dilatation, and expansion of the skull thus induced will depend directly upon the youth of the infant or child. As a rule, secondary hydrocephalus never reaches the extent of the primary form, owing to the rapidly fatal nature of its cause. In these cases we seldom see pressure effects beyond flattening of the convolutions and moderate expansion of the cranial vault.

The cases of acute hydrocephalus due to meningitis serosa, and the cases in which a defect of brain substance is counterbalanced by an equal bulk of cerebro-spinal fluid, do not commonly fall under the heading of this paper.

In chronic hydrocephalus internus there seems to be a special susceptibility of the membranes to acute disease, so that at autopsy it is not uncommon to find

evidence of an acute meningitis, simple, hæmorrhagic, suppurative, or tubercular.

The fluid found in hydrocephalic idiots has been frequently analyzed. In a case of Bourneville's the analysis of the hydrocephalic fluid, withdrawn nine hours after death, resulted as follows: Color, pale yellow; aspect, clear after standing; reaction, neutral; odor, like that of blood; consistence, slightly viscous; density, 1.006; organic matter, 1.65; salts, 10; total fixed solids, 11.65; phosphoric acid, 0.22; sodium chloride, 0.80; albumin, 0.26; leucocytes, very few; red blood-corpuscles, considerable.

SYMPTOMATOLOGY.—The enlargement of the head is ordinarily the first symptom to attract attention. It

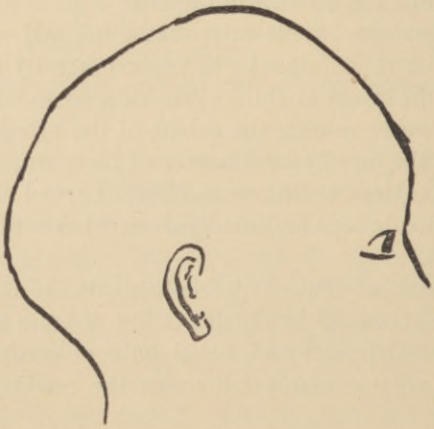

FIG. 2.—The naso-occipital arc in oxy-hydrocephalus.

may be so large at birth as to interfere with delivery, or the expansion may not be noteworthy until during

the first months or first years of life. While there is commonly a great similarity of cephalic contour in most cases, so that we have come to look upon a certain slope of head as a characteristic type, this is by no means always true, for irregular synostoses may alter the outlines considerably (see Figs, 2, 3, 4, and 5).

In a typical hydrocephalic child the head is quite evenly expanded into a somewhat globular shape, when

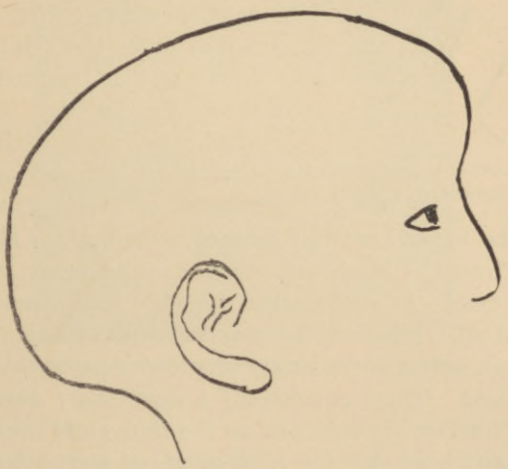

Fig. 3.—The naso-occipital arc in scapho-hydrocephalus.

viewed from the lateral aspect (Fig. 6). Looking down upon the outline of the horizontal circumference, it has rather a triangular contour with the apex forward (trigonocephalus). The one illustrated in Fig. 5 has a perfectly round head. The face seems puny and weazened in contrast to the enormous head above it, and is triangular

in shape. In young hydrocephalic patients the fontanelles gape widely, and the sutures are often open, so that the pulsations synchronous with the pulse, the hydrocephalic souffle, and fluctuation may be perceptible. In older cases both sutures and fontanelles are commonly closed. Examination with electric light, either in the throat or at the side of the head, will in marked cases show translucency of the skull. Young children, through weakness of the muscles of the neck, tend to let the heavy head fall forward, backward, or sidewise. Later on the head, even though quite heavy, may be held erect. In very marked cases the head is never held up at all. The forehead may be wrinkled as if in pain, though ordinarily in slowly progressive patients the face is placid. There is generally a superciliary depression. The features are pale, and on the thin white skin of the face and head the blue veins stand out markedly. Dentition is commonly retarded, and the teeth are badly preserved, though sometimes they are exceptionally fine. Squint and nystagmus are not infrequent.

FIG. 4.—The naso-occipital arc in another form of oxy-hydrocephalus.

GENERAL PHYSICAL CONDITION.—Rhachitic deformities of the thorax are not uncommon, such as the thoracic chaplet, exaggeration of the curvature of the posterior angle of the ribs, and slight kyphosis or scoliosis, or both. The abdomen is frequently enlarged. The pelvis is seldom affected except in rhachitic cases, where its altered

position is purely compensatory. There is general muscular weakness. Anæmia and emaciation are common,

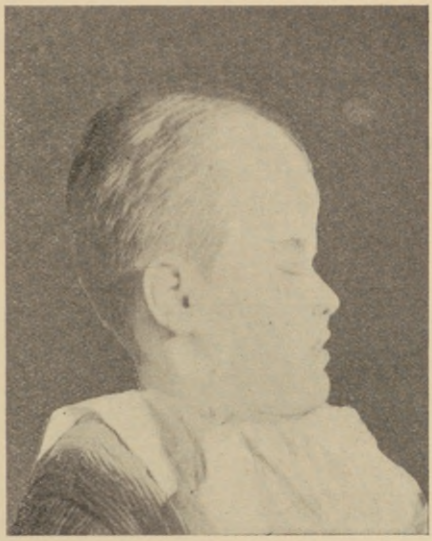

FIG. 5.—An oxy-hydrocephalic idiot at Randall's Island.

and nutrition, as a rule, is so impaired that a great number have a sort of progressive cachexia or marasmus. They usually die of pulmonary congestion. There are exceptional cases, however, in which the general health is excellent. Subnormal bodily temperature is the rule in hydrocephalus. Increase in weight and bodily growth often proceed as in normal children, except in cases presenting paralyses. Puberty is apt to be retarded in bad cases. Learning to walk is retarded considerably, even if no paralysis be present, when the disease begins early. The gait is apt to be affected by the difficulty of balanc-

ing the head, so that it is slow, shambling, weak, and awkward to a degree depending upon the extent of the interference.

Paralysis.—Very often we meet with paralyses which differ in degree and character as much as do the infantile cerebral palsies in general. They depend usually upon pressure-atrophy of the cerebral segments of the motor tracts. Hence their development is slow—first a paresis, then complete paralysis; we meet with monoplegias, hemiplegias, paraplegias, and diplegias. As a result, we find in the paralytic limbs retardation of growth, ex-

Fig. 6.—Three cases of hydrocephalic idiocy showing typical head contours.

aggerated reflexes, and spastic contractures. Hemiplegia naturally signifies a greater increase of ventricular fluid on one side than on the other, or suggests dilatation of one ventricle alone. I have not observed in this species of infantile cerebral palsy the methemiplegic movements common to other forms. When hydrocephalus is suspected of being secondary to cerebellar neoplasms, the motor disorders characteristic of such lesions should be looked for.

Epileptoid Convulsions.—Convulsions not only generally usher in the last stages of the disorder, but they are not at all infrequent during its progress, constituting sometimes a real symptomatic epilepsy. Occasionally the epilepsy is local—a hemi-epilepsy—but general convulsions are the rule. While we are disposed, as a rule, to look upon such explosions as cortical in their origin, episodes significant of critical increases of cortical pressure, this is not always true. My case, reported above, of a hydrocephalic child with scarcely any cortex left in its wasted brain (portions, too, of only the temporo-occipital cortex) was further remarkable in that it had occasional convulsions, the last at the time of its death.

General and Special Sensibility.—Cutaneous sensibility is rarely affected, except in extreme cases where the sensory tracts are involved. Taste, hearing, and smell likewise seldom suffer. Enfeeblement of vision and great myopia are constant symptoms, and total blindness is quite common. In primary hydrocephalus there is rarely any pain or headache.

Psychology.—The mental symptoms in hydrocephalic idiocy vary greatly according to the degree of injury to the intellectual substratum. The tendency is to progressive enfeeblement along all lines of psychic growth, so that we find among them each degree of impairment, from feeble-mindedness to imbecility and complete idiocy. Where the malady is considerable, the patient finds it difficult to support the head, and, consequently, inclines to lie in bed or to sit still with the head hanging down. He gradually loses any play of expression he may have had. The sad, dejected look is commonly the last expression to take its departure.

A profound apathy is noticeable in looks, attitudes, and movements. The faculty of attention gradually loses its sharpness and grows more and more unstable.

In the early stages he has all of the normal instincts, but with the evolution of the disorder these fade away, and the patient enters upon a purely vegetative existence, with a tendency to sleep and to speak little if he speak at all. The hydrocephalic idiot is ordinarily timid, fearful, gentle, neither affectionate nor mischievous, with a certain refinement of face, expressive of sadness and languor. Sollier looks upon the hydrocephalic idiot as a complete contrast to the microcephalic. Language is apt to be slowly developed, and later slowly lost. The will is almost absent as a rule. Memory is at first fair, then gradually impaired, and, finally, vanishes wholly. A peculiarity in a considerable number of cases of hydrocephalus is a tendency to sudden accesses of anger, which Bourneville very suggestively compares to the convulsive crises of the malady. Co-ordinated tics, such as antero-posterior and lateral oscillation, are sometimes met with in this as in other forms of idiocy. The sentiment of vanity seems to be rather strong, especially among hydrocephalic females. While the mental faculties are generally much impaired in all directions in severe cases, distinctive psychological features can not be portrayed in connection with hydrocephalic idiots, and it is often remarkable how much intelligence and expression may remain in an individual apparently afflicted to the extremest degree.

Course and Duration.—Nothing is more uncertain than the course of hydrocephalus. Sometimes beginning before birth, the process may cease for a long period or altogether. In the greater number of

cases it is a few months after birth before the increase of fluid begins to manifest itself, the infant up to this time developing like normal children. Then convulsive crises or meningitic congestions announce a new inception of the malady. Spontaneous recoveries or arrests are not infrequent. On the other hand, the progress may be steady and continuous and slow or rapid. The inception of the disorder may be so insidious and the course so protracted that it is only discovered by accident (the misfit of a cap or hat) that anything is wrong. Some idea of the varying progress of cases may be gathered from the following cephalic measurements taken from Bourneville's reports:

Case.	I.		II.		VI.			XVI.		XXI.	
Year.	1886	1887	1886	1889	1887 Apr.	1887 Jun.		1880	1893	1886	1893
Circumference in centimetres.	58·0	59·5	60·0	62·5	46·0	51·0		53·0	57·0	36·0	60·2
Binauricular arc...	34·0	39·0	42·0	46·5	32·5	34·5		37·0	38·0	37·0	42·0
Naso-occipital arc.	34·0	35·5	42·0	44·5	30·5	37·0		38·0	41·5	35·0	41·0
Antero - posterior diameter.	19·5	19·0	19·8	20·5	15·5	16·8		18·0	20·3	19·3	20·2
Binauricular diameter.	12·7	12 5	18·0	20·0	11·0	11·0		10·5	12·0	18·0	17·8
Biparietal diameter	16·0	16·5	18·0	18·6	14·2	14·5		14·0	15·0	13·5	14·6

Rokitansky reports a case of enormous hydrocephalus where spontaneous rupture through one of the sutures took place with the result of ultimate recovery. The common termination of the disease is from bronchopneumonia or meningitis at the average age of three to six years.

Diagnosis.—It is not often that there is any difficulty in diagnosticating the affection when progressive enlargement of the head, with separation of the sutures and thinning of the cranial bones, is apparent. There are

cases of cerebral hypertrophy and of hyperostosis of the cranial bones which may at times be confused with chronic hydrocephalus, though it must be remembered that the meningitic and convulsive crises of the latter are distinctive. The most puzzling cases are those of hydrocephalus acquired after union of the sutures. These are, however, rare cases, and, as a rule, symptomatic of other cerebral affections. In hydrocephalus which appears to be symptomatic, headache and vomiting, absent in primary hydrocephalus, should suggest neoplasm as a cause. Lumbar puncture for the withdrawal and examination of the cerebro-spinal fluid for bacilli should be resorted to whenever the question of tuberculous meningitis comes up as a question of diagnosis.

The ophthalmoscope should be employed to determine neuritis and atrophy of the optic nerve, not found in cerebral hypertrophy or cranial hyperostosis, but not infrequently in hydrocephalus.

Prognosis.—Naturally, the prognosis is grave. Death in childhood from some intercurrent affection is the usual termination, though spontaneous arrest of the progress of the disease, with a fairly long life subsequently, is known to occur at times. Complete recovery is, of course, extremely rare, for the damage done to the brain by retardation of its development or by abrogation of some of its functions can be only faultily repaired. If much harm has already been done, such as the onset of paralysis of a limb or limbs, or the production of blindness, the probability of an individual's survival beyond childhood is small. Bourneville quotes Gratiolet as believing that a certain degree of hydrocephalus, by dilating the cranial cavity, may favor the development

of the brain, and thus give rise to intellectual growth of a high order, as instanced in the case of Cuvier. It is certainly true that in our social life we occasionally meet with individuals of intellectual superiority whose cephalic contours would indicate an arrested chronic hydrocephalus. Thackeray has been cited as an instance of genius coexisting with hydrocephalus. However this may be, I am inclined to agree with Bourneville that the skull is an elastic vestment of the brain depending for its growth altogether upon the development of the encephalon.

TREATMENT.—The treatment of chronic hydrocephalus of the simple, primary kind is medical or surgical. Bourneville sums up the medical treatment, which he has found successful in some instances, as follows:

1. Compression (Barnard, Trousseau).
2. Revulsives (Gelis).
3. Internal administration of calomel (Gelis).

The head of the child is shaved and a capeline bandage is kept applied for a week. After a week's rest, it is renewed, or, if any incident contraindicates this, daily friction with mercurial ointment is substituted. At the same time calomel is administered twice weekly, in one-and-a-half-grain doses. Every month, for a year or more, a vesicatory is applied to the head for from fifteen to twenty hours, and when it begins to dry compression is again employed. In addition to this, exercise, massage of the limbs, salt baths, douches, tonics, etc., are used as required. Pedagogic treatment is important for this class of cases when cure or arrest of the disorders seems evident.

Thus far surgical procedure of any kind has not proved to be of particular value. Most patients have died

as a result of operation. Possibly a more conservative method of operating may be devised in the future. Up to the present time the surgical procedures resorted to have been (1) craniectomy and drainage; (2) puncture of the lateral ventricles; (3) puncture of the fourth ventricle; (4) lumbar puncture.

Craniectomy with drainage may be excluded nowadays altogether from the category of expedients. The patient is almost certain to die as a result of the sudden evacuation of a large amount of fluid.

Puncture of the lateral ventricles, even if carried out carefully and skillfully according to Keen's method (*Medical News*, December 1, 1888), promises but little more, and had best be abandoned as a remedial measure in this class of cases.

Trephining of the occipital bone, three fourths of an inch below the superior curved line, to the right of the middle line, with subsequent enlargement of the opening downward, and the insertion of a probe into the fourth ventricle, is an operation which was proposed by Parkin (*Lancet*, 1893) and carried out in four cases with the recovery of two. It seems to me that on theoretical grounds this form of procedure has much in its favor, and it deserves more extensive trial. Another similar measure, successfully tried, is trephining in the region of the temple and insertion of the cannula in the subarachnoid space of the fissure of Sylvius.

Lumbar puncture, by which is meant the insertion of a drainage needle through an intervertebral space in the lumbar portion of the spinal column into the spinal cavity, has been rather extensively employed of late in a great variety of affections of the central nervous system, hydrocephalus being among them. Jacoby (*New York Medical Journal*, December 28, 1895, and January 4,

1896) has recently made a careful study of the subject of lumbar puncture in connection with a number of central nervous maladies. Quincke seems to have been the originator of the method (Tenth International Congress, 1891), though Wynter (*Lancet*, 1891) proposed it about the same time. It seems to be a simple and harmless procedure, as well as a powerful means of reducing intracranial pressure. An ordinary aspirating needle (a millimetre in diameter, eight centimetres long) is aseptically employed. The space between the third and fourth lumbar vertebræ is selected, five millimetres to the right of the median line, though in children the puncture may be made directly in the median line. An anæsthetic is rarely required. The needle should be pushed in for from two to eight centimetres. The fluid begins to flow out in drops or in a small stream according to the degree of intracranial pressure. The amount of fluid allowed to flow out must be gauged by the indications and conditions. Fürbringer withdrew in one case a hundred and ten cubic centimetres at one time. On removing the needle, the puncture is closed with a little iodoform collodion and a compress with adhesive plaster. Fürbringer, Heubner, and Ewald report temporary improvement in hydrocephalus after lumbar puncture, but the method is as yet too novel and has been too little used to allow of any definite conclusions as to its value in this class of cases. Several cases of recovery from hydrocephalus by means of this measure have been reported.

Operation does not preclude the employment of other means also. Horizontal bandaging of the head with a wide rubber bandage after tapping, and the use of cod-liver oil and phosphorus, are certainly of considerable service, and should not be neglected.

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