HYDROCEPHALUS.

BY JOHN FRASER AND NORMAN M. DOTT, EDINBURGH.

HYDROCEPHALUS has hitherto ranked as one of the most intractable and unpromising of the diseases of childhood. A multitude of surgical procedures has from time to time been instituted for the relief of the condition, but almost without exception they have failed in their purpose, and the rare occasions on which they have succeeded have left one wondering whether, after all, the success was no more than a coincidence—whether, in fact, improvement would have occurred if nothing active had been done, for spontaneous arrest of hydrocephalus is an actual—though rare—possibility. This unsatisfactory state of affairs depended largely upon erroneous ideas regarding the etiological pathology of the disease.

As early as 1862 Hilton,¹ in his *Lectures on Rest and Pain*, described varieties of obstructive hydrocephalus arising from congenital obliteration of the aqueduct of Sylvius, and from occlusion of the foramina present in the roof of the fourth ventricle. The clue which Hilton thus gave to the pathology of certain types of the disease does not appear to have produced in the past any operative interference of lasting and definite value, and we owe to Dandy and his co-workers the establishment of operative principles which hold out the prospect, as far as we can at present tell, of permanent cure of the disease.

In the course of our work we have had the opportunity of treating a number of children and infants, the victims of hydrocephalus, and since the publication of Dandy's² original papers we have followed up the subject with the closest interest; after four years partly spent in observing and treating these cases we venture to put forward our experiences and our results.

THE NORMAL ANATOMY AND PHYSIOLOGY OF THE PARTS INVOLVED.]

It is essential to have a proper idea of the processes involved in the production and absorption of the cerebrospinal fluid, and of the relationship of the parts connected with its circulation. Such knowledge is fundamental, because the development of hydrocephalus necessarily entails an error either in the production, circulation, or absorption of the fluid.

The Production of the Cerebrospinal Fluid.—

1. *From the Choroid Plexuses.*—We may assume that the bulk of the cerebrospinal fluid is derived from the choroid plexuses which lie in the lateral ventricles. Dandy³ has produced confirmatory evidence of this assumption by experiments on dogs. He occluded the foramen of Monro by a strip of fascia or peritoneum, and a dilatation of the corresponding lateral ventricle resulted. He occluded the iter by means of an oiled gelatin capsule containing cotton-wool, and a dilatation of the two lateral and the third ventricles ensued. He repeated these experiments, excising in the first case the corresponding choroid plexus, and in the second the choroid plexus of both lateral ventricles. In the first instance the lateral ventricle collapsed completely; in the second, a slight degree of dilatation of the ventricles occurred on account of the remaining choroid plexus of the third ventricle. Absolute proof was thus given that the choroid plexus is the site of production of the cerebrospinal fluid.

The method by which the fluid is actually produced from the villi is uncertain. It is not yet possible to say whether the process is one of secretion, filtration, or dialysation. The fluid differs so widely from that secreted by most other glands, and it so persistently retains its freedom from contamination by such body fluids as bile in jaundice, or by drugs

when they are injected into the body, that we must assume that whatever the method of production, certain strong selective actions are at work. The time in embryonic life at which the formation of the fluid first appears remains doubtful, but one anatomical detail would suggest that it is unlikely it is produced in any quantity before the fifth month of intra-uterine life. It is only after the fifth month that the tela choroidea demonstrates the perforations of the foramina of Magendie and Luschka. Therefore any production in quantity of cerebrospinal fluid before the fifth month would have no obvious means of exit from the ventricular system.

2. *From the Perivascular Spaces.*—Another possible source of the production of the cerebrospinal fluid exists in the perivascular spaces. Lymphatics, as we generally understand them, do not exist in the brain or meninges; their place is taken by perivascular spaces or channels, and these cannot be grouped as lymphatics, because the fluid which they contain has neither the constitution nor the characters of lymph. The perivascular

FIG. 130.—Sagittal section through head, showing the anatomy of the ventricular system and basal subarachnoid cisterns. (A) Cisterna interpeduncularis. (B) Septum pellucidum. (C) Foramen of Monro. (D) Choroid plexus of third ventricle. (E) Third ventricle. (F) Aqueduct of Sylvius. (G) Fourth ventricle. (H) Choroid plexus of fourth ventricle. (J) Foramen of Magendie. (K) Cisterna magna. (L) Central canal of spinal cord.

spaces extend throughout the cerebral vascular system, even to the finer capillaries (their presence was fully demonstrated by Spina⁴), and it would seem that they contain fluid of simpler constitution than lymph, which passes into them from the blood-stream.

The perivascular spaces communicate with the subarachnoid space, and therefore the fluid of the former must to some extent be included in the cerebrospinal fluid. It is an interesting point in the physiology of the central nervous system, though not directly applicable to the subject of hydrocephalus, that the waste products of the central nervous system probably accumulate in the fluid of the perivascular spaces, and are thence conveyed into the subarachnoid space to mix with the cerebrospinal fluid.

The Course of the Fluid after Production.—The fluid circulates along the ventricular system and escapes from it through the medium of the central rounded foramen

of Magendie, and the more slit-like lateral foramina of Luschka. It is now in the subarachnoid space, and at first it tends to collect in the large subarachnoid cisterns which lie at the base of the brain (*Fig. 130*), the cisterna magna on the dorsal aspect and the cisterna pontis with its various subdivisions on the ventral aspect. The value of the cisterns is two-fold—they stabilize the pressure of the cerebrospinal fluid, and they render possible a uniform distribution of the fluid in its further progress over the cerebral hemispheres. From the subarachnoid spaces and cisterns the fluid passes in two directions—a small proportion passes downwards through the foramen magnum into the subarachnoid space of the spinal meninges, while the larger proportion escapes upwards between the mid-brain and the tentorium cerebelli to be distributed in the wide area of the subarachnoid space which covers the cerebral hemispheres: it is in this area that absorption of the fluid occurs.

The Method of Absorption of the Fluid.—

1. *Pacchionian Bodies*.—It was formerly held that the Pacchionian bodies were the media through which the cerebrospinal fluid passed from the subarachnoid spaces into the cerebral sinuses (*Fig. 131*). Recently, however, objection has been raised to this view for two reasons—first, because the Pacchionian bodies are demonstrable only in man and in certain anthropoids; secondly, because in man and anthropoids they are only met with in adult life.

2. *Arachnoid Villi*.—Weed⁵ and his collaborators have demonstrated that, while the Pacchionian bodies undoubtedly play a part in absorption of the cerebrospinal fluid, other means exist. Using a Prussian-blue reaction they were able to demonstrate the occurrence of what they have termed 'arachnoid villi'. These are delicate coil-like structures of interlacing strands of connective tissue prolonged from the arachnoid into the walls of the dural sinuses. The Pacchionian bodies are examples of hypertrophied arachnoid villi, and this explanation disposes of the two objections already quoted. There is, therefore, the passage of the cerebrospinal fluid into the bloodstream of the dural sinuses through the medium of the arachnoid villi and, in later life, through hypertrophied examples of these—the Pacchionian bodies. The means by which the cerebrospinal fluid passes from the arachnoid villi into the venous sinuses is a double one—there is a process of filtration from a point of higher to one of lower pressure, and there is a process of osmosis from a fluid of low colloid and crystalline content to one of higher content.

3. *Arachnoid Mesothelial Cells*.—Weed⁶ has demonstrated another method by which absorption of the cerebrospinal fluid may occur. Surgeons have recognized that on exposing the outer aspect of the dura (as, for example, in such a procedure as an osteoplastic craniotomy) there is often a faint oozing of cerebrospinal fluid through what appear to be tiny pores in the dural structure: further, the blood in this situation has a distinctive watery appearance. Weed believes he has explained these occurrences by demonstrating small nest-like collections of arachnoid mesothelial cells which lie in the dura and are in continuity with the arachnoid villi. Through such channels there is in all probability a slight escape of cerebrospinal fluid on to the outer surface of the dura, where it is absorbed into the circulation.

FIG. 131.—Diagrammatic representation of mechanism of absorption of cerebrospinal fluid, showing arachnoid villi connected with subarachnoid space and projecting into a cerebral blood sinus. (A) Subarachnoid space. (B) Lacuna lateralis. (C) Superior longitudinal sinus. (D) Lacuna lateralis. (E) Subarachnoid space. (F) Subarachnoid space. (G) Falx cerebri. (H, J) Arachnoid villi (Pacchionian bodies).

4. *Lymphatic Absorption.*—An accessory pathway of absorption exists through the medium of the lymphatic system. Key and Retzius were able to inject the cervical lymphatics from the spinal subarachnoid space, and later observers have obtained similar results. The practical importance of this demonstration was not fully appreciated until Weed, using the Prussian-blue method, showed that a subarachnoid injection reached the perineural lymphatics of the cranial nerves and the cervical vessels and glands. Evidently, then, the subarachnoid space is continuous along the perineural spaces of the cranial nerves with the perineural lymphatic channels, and through the medium of this connection an absorption of cerebrospinal fluid must occur.

The Special Anatomy Involved.—Of the special anatomy of the parts little need be said. A description of many of the anatomical details is unnecessary in a contribution of this description; therefore only those points which have a direct bearing on the problems of hydrocephalus are mentioned.

The large proportion of the cerebrospinal fluid is formed within the lateral ventricles in which lies the bulk of the choroid plexus. The fluid passes by the foramina of Monro into the narrow cleft-like space which separates the mesial surfaces of the thalami—the third ventricle. This ventricle contains a choroid plexus, and therefore it is the site of production of a certain amount of cerebrospinal fluid.

The fluid leaves the third ventricle by the Sylvian aqueduct to pass into the fourth ventricle. The lumen of the Sylvian aqueduct is somewhat narrowed at its two extremities—a point of importance we shall allude to later in connection with congenital obstructive hydrocephalus. In regard to the fourth ventricle, there are certain details in the roof of the ventricle which require mention, as they have a close bearing on the subject under discussion. Viewed in median sagittal section (see *Fig. 130*), the roof of the ventricle appears as a tent-like structure, the wings of which, where they come together, bound the space 'the recessus tecti', which penetrates the cerebellar medulla between the superior and inferior vermes. The upper wing of the tent is formed by the 'superior medullary velum' connected with the corpora quadrigemina above, the cerebellar medulla below and the superior cerebellar peduncles laterally.

It is, however, in the lower wing of the tent that, from our point of view, the chief interest lies (*Fig. 132*). It is composed of two parts, an upper thicker crescentic plate of white matter (the inferior medullary velum) and a lower extremely thin membrane (the tela choroidea). The latter structure is formed in a morphological sense of ependyma only, though actually it is supported by a backing of pial tissue. During the early part of foetal life the tela choroidea is a complete membrane, but about the fifth month it becomes perforated at its lower extremity by an aperture which remains throughout life, the foramen of Magendie. About the same time two additional clefts (the foramina of Luschka) appear at the lateral extremities of the tela, behind the upper roots of the ninth nerve in the pouch-like extension of the ventricle beneath the flocculus, and through these three apertures, and probably through them alone, the system of ventricular cavities and the central canal of the spinal cord are brought into communication with the subarachnoid space.

FIG. 132.—Dissection exposing the roof of the fourth ventricle. The lower part of the roof has been partly opened and turned down. (A) Lamina quadrigemina. (B) Roof of fourth ventricle. (C) Choroid plexus of fourth ventricle. (D) Third ventricle. (E) Pineal body. (F) Floor of fourth ventricle. (G) Foramen of Luschka. (H) Foramen of Magendie. (J) Medulla oblongata.

Three groups of choroid plexus appear in the roof of the fourth ventricle—a medial and two lateral—and therefore a proportion of the cerebrospinal fluid is formed within this space.

Having escaped from the fourth ventricle into the subarachnoid space by the foramina of Magendie and Luschka, the fluid is free to pass forwards over the hemispheres or caudally into the spinal subarachnoid space. We have elsewhere described the absorption of the fluid from the cranial subarachnoid space, and we are therefore now in a position to appreciate the cycle of the fluid from its point of production to the area of its resorption into the blood-stream of the various sinuses.

THE ESSENTIAL ERROR IN THE FORMATION OF A HYDROCEPHALUS.

By the term hydrocephalus we mean an accumulation of cerebrospinal fluid within the cavity of the skull, and, keeping in view the outline which we have given of the formation, circulation, and absorption of the cerebrospinal fluid, we may systematize the possible origins of the disease as follows : (a) *It may be due to an excessive production of fluid ;* (b) *It may be due to an obstruction in some portion of the route along which the fluid circulates ;* (c) *The production and circulation of the fluid remaining normal, there may be an interference with the absorption of the fluid.*

Based upon these three possibilities, a reasonable and scientific classification of the disease can be made, which will include moreover all varieties of the disease.

VARIETIES OF CLASSIFICATION.

Hitherto different varieties of hydrocephalus were recognized in describing the disease. For example, hydrocephalus was classified as 'acute' or 'chronic', according to the rapidity with which the fluid accumulated, or dependent on its association with coincident acute inflammatory reaction in the meningeal structures. Hydrocephalus was spoken of as 'internal' or 'external', according to whether the fluid accumulated within the ventricular system or in the extracerebral tissue of the arachnoid spaces. Further, the disease was classified as 'congenital' or 'acquired', according to the history of its incidence and occurrence. Such an irregular and rambling classification had no proper basis. The occurrence of an external hydrocephalus as a primary condition distinct from a co-existing internal hydrocephalus has been questioned, and its occurrence is universally accepted as a very rare condition. In our personal experience we have seen two examples of it. Both were cases in which a marked degree of hydrocephalus was present at birth, and in both cases life persisted for only a few hours after birth. In the two instances a similar condition was found (*Fig. 133*) ; there was a congenital absence of the quadrigeminal plate and the superior cerebellar peduncles.

FIG. 133.—Drawing from a specimen of congenital (developmental) hydrocephalus. The brain and skull are grossly deformed. The upper part of the quadrigeminal plate is absent, and the open ends of the Sylvian aqueduct are seen. The cerebral tissue forming the roofs of the lateral ventricles is defective, so that these cavities open on to the dorsal surface of the brain. A ruptured membrane of attenuated cerebral tissue attached to the right defect suggests an extreme ventricular distention at an earlier period. The posterior fossa of the skull is disproportionately small and shallow.

The posterior wall of the Sylvian aqueduct was absent, and the cerebrospinal fluid which was produced within the ventricles was poured directly into the sub-meningeal space. The result was that the cerebral tissues were atrophied and ill-developed, and displaced forwards into the frontal regions of the cranium, the greater portion of the skull being occupied by cerebrospinal fluid. The condition, therefore, approximated as closely as possible to a pure external hydrocephalus; but, even in this instance, there was to some extent a co-existing, though slight, internal hydrocephalus.

The classification of 'acute' and 'chronic' hydrocephalus was specially unfortunate. Apparently in the minds of many observers acute hydrocephalus was synonymous with the development of a basal meningitis. But, whatever the origin of the condition, all varieties of hydrocephalus are chronic in their formation, though the actual period of time involved may vary within wide limits.

Something is to be said for the method of classification into 'congenital' and 'acquired' varieties of the disease—a considerable proportion of cases can without much difficulty be grouped into one or other of these two classes—but the independent use of the two terms is not sufficiently descriptive of the condition of affairs.

The Classification Suggested by Dandy.—Recognizing the unsatisfactory condition of the classification nomenclature, Dandy⁷ has suggested that the following is a scientific and inclusive grouping of the possibilities :—

Hydrocephalus due to	Diminished absorption of cerebro-spinal fluid	<ol style="list-style-type: none"> 1. Communicating hydrocephalus (due to adhesions in the subarachnoid space) 2. Obstructive hydrocephalus— Congenital, atresia, adhesions, tumours 3. External hydrocephalus?
	Increased production of cerebro-spinal fluid	<ol style="list-style-type: none"> 1. Acute hydrocephalus (increased fluid from inflammatory products in acute meningitis and trauma) 2. Communicating hydrocephalus (due to occlusions of the vena magna Galeni)

This classification is complete in so far as it would appear to include all varieties of hydrocephalus. From the point of view of the pediatric surgeon, however, it includes types of the disease which, if they actually exist as clinical entities, have little or no relationship to surgical treatment. Under this criticism we include external hydrocephalus, acute hydrocephalus, and communicating hydrocephalus due to occlusion of the vein of Galen.

We have stated our experience with external hydrocephalus; we do not believe that acute hydrocephalus exists as a condition calling for surgical interference; and, while we realize that a non-obstructive variety of hydrocephalus, said to be due to occlusion of the vein of Galen, has been produced experimentally (Dandy⁸), its clinical existence is exceedingly doubtful, and it certainly can be accepted as negligible in our estimation of hydrocephalus work.

Excluding, therefore, for practical purposes these three varieties, only the congenital anomalies and communicating and obstructive types (Dandy) remain to be considered.

We would further venture to criticize the title of 'communicating hydrocephalus' (as contrasted with 'obstructive hydrocephalus') as being insufficiently expressive of the condition, or even misleading. According to Dandy, the 'communicating' variety depends upon pia-arachnoid adhesions. The normal filmy pia-arachnoid tissue is replaced by a firm, fibrous, adherent membrane; and further, it is the distribution and location of these adhesions, not their extent, which determine the production of hydrocephalus. Adhesions encircling the mid-brain where it passes through the incisura tentorii interrupt the subarachnoid communication between the posterior and middle cranial fossæ, and thereby eliminate the cerebral subarachnoid space (the main area of absorption) from participation in the absorption of the fluid. Adhesions which obliterate the cisterna magna or those at the base of the brain will produce hydrocephalus as effectively. If this reasoning is correct, 'communicating hydrocephalus' is as much an example of an obstructive lesion as the intracerebral types of obstruction.

Returning, then, to our original conception of the possible causes of the disease, our experience leads us to the conclusion that increased formation of fluid may be excluded as of little practical importance, and that deficient absorption is not known to occur *per se*. Therefore, for practical purposes, we are only concerned with hydrocephalus which is due to congenital anomalies or to an obstruction to the circulation of the cerebro-spinal fluid. The latter type is naturally divided into two main groups in which the obstructive lesion is ventricular or extraventricular. The ventricular group is again divided into degrees according to the exact anatomical site of the lesion.

A More Suitable Classification.—On the grounds above stated, we suggest the following as a more suitable classification:—

GROUP		DEGREE
		(Site of obstruction)
Hydrocephalus due to	Congenital anomalies	
	Ventricular (obstruction)	<ol style="list-style-type: none"> 1. Between one lateral and 3rd ventricle 2. Between both lateral ventricles and 3rd ventricle 3. Between 3rd and 4th ventricles 4. In the roof of 4th ventricle
	Extraventricular (obstruction)	In the subarachnoid space

The above, we submit, is a simple, expressive, and accurate classification from the anatomical and physiological points of view. It is also very significant practically, for, as will appear later, each type has its distinctive and appropriate treatment.

In actual use we omit the term 'obstructive' entirely, and we speak, for example, of a ventricular hydrocephalus of the fourth degree, or of an extraventricular hydrocephalus.

THE ETIOLOGICAL PATHOLOGY OF HYDROCEPHALUS.

It is important to explain that the conclusions arrived at have been based entirely upon the observations made on the 21 cases under review, with the addition of certain other cases which, through the kindness of our colleagues, we have had an opportunity of studying.

1. Congenital Hydrocephalus and the Conditions under which it is met.—Under two varieties of conditions hydrocephalus has been met with at birth, and therefore the varieties may be described as examples of congenital types.

a. True Congenital Hydrocephalus.—There is a well-developed form in which the head is considerably enlarged at birth, so much so, sometimes, as to constitute an obstruction to delivery. This variety we have found to be associated with an error in the development of the central nervous system, which permits an escape of fluid from an open Sylvian aqueduct. The resulting hydrocephalus is of an external character, though there may be an associated dilatation of the ventricular spaces. It is not amenable to surgical treatment, and it is rarely compatible with post-natal life for any prolonged period.

b. The Variety of Hydrocephalus which Accompanies a Spina Bifida.—At birth attention is naturally directed to the spinal error, and the head condition may be overlooked, but we have satisfied ourselves that a certain proportion of spina bifida cases are accompanied by examples of congenital hydrocephalus inasmuch as there is a ventricular dilatation present at birth; we have confirmed this observation by ventriculography during life, and by post-mortem examination. If the spina bifida is operated on and the defect closed, the pre-existing hydrocephalus soon forces itself on one's attention by the rapid increase in the size of the head. This variety of hydrocephalus is of the extraventricular type in so far as there is no obstruction to the circulation of the fluid up to the point of its exit from the fourth ventricle.

These varieties of hydrocephalus (*a* and *b*) are the only ones which we believe to be congenital, and both of them are closely related to an error in the development of the central nervous system.

2. The Origin of Ventricular Hydrocephalus.—

a. Tumours as a Cause.—In a class by itself comes that group which owes the development of the hydrocephalus to the presence of a cerebral tumour. Theoretically a tumour which from its position presses upon any portion of the interventricular system may give rise to a hydrocephalus; practically, it is the cerebellopontine angle which is the usual situation of the neoplasm (*Fig. 134*). Its pressure is exerted upon the lower portion of the

FIG. 134.—Drawing from a specimen from a case of secondary hydrocephalus. The condition developed secondarily to a cerebral tumour which occluded the lower part of the Sylvian duct, thus leading to a ventricular hydrocephalus of the third degree.

Sylvian aqueduct, and there is a resulting dilatation of the ventricular system above this level. Though in a very real way the hydrocephalus is a secondary development to the tumour, it is interesting that, in the examples of this condition which came under our care, the hydrocephalic condition clinically overshadowed the local tumour formation, and the clinical features which impressed themselves on the examiner were those of the ventricular hydrocephalus rather than the signs of the neoplastic condition.

As we have said, this type of obstructive hydrocephalus stands in a group by itself, and surgical interference is primarily directed towards the tumour; it does not rank as a primary hydrocephalus, and therefore it does not enter into a contribution of this description. There remains a large group of cases of ventricular hydrocephalus for which other causes must be found. Atresiae and adhesions constitute the remaining causal factors.

By the term 'atresia' we mean a narrowing or obliteration of a portion of the interventricular system, while by the use of the term 'adhesions' we imply that adhesions have formed at the base of the brain so as either to occlude the foramina in the roof of the fourth ventricle or to limit the circulation of the fluid in its immediate neighbourhood. From the etiological point of view both of these types have a common basis in so far as both varieties have an inflammatory origin, plastic or infective. We have not met with an atresia which could be accepted as an actual congenital error in the cerebral development—this is an important consideration. There is a wide variety of factors which may lead to the formation of adhesions, and certain causes are apparently more liable than others to produce the errors.

b. Birth Hæmorrhage as a Cause.—

When investigating the case-histories it may be found that the birth was a difficult one, that forceps were employed, and that a good deal of trouble was experienced in the delivery of the head. In fact, in certain cases there was the likelihood that an intracranial hæmorrhage had occurred. In seven cases of the series under consideration a natal history of this sort was obtained, and the subsequent operative interference revealed the presence of a hæmorrhagic effusion in the membranes at the base of the brain; in certain of these the hæmorrhagic effusion was definitely invading the roof of the fourth ventricle, and it could be demonstrated that there was a resulting occlusion of the foramina normally present in the ventricle roof (*Fig. 135*). It would appear that a basal hæmorrhage of this description is more likely to occur when the forceps blades are applied in the sagittal axis of the head than in the coronal axis. It seems reasonable that the compression of a forceps blade beneath the occiput is more likely to produce a subtentorial hæmorrhage than forceps applied laterally.

We possess very definite evidence that an intracranial hæmorrhage produced at birth is liable to be followed by a hydrocephalus if the location of the hæmorrhage is subtentorial in its distribution. These remarks apply at this stage only to ventricular hydrocephalus, but we shall allude to the subject again in relation to the extraventricular type.

*c. Infective Meningeal Conditions as a Cause.—*It is possible that we underestimate the degree of occurrence of infective meningeal conditions in the early period of life: the imperfect ossification of the base of the skull may explain a greater liability to infection from the extracranial regions at this stage than in later life. At any rate, during infancy there is a definite proportion of children who suffer from inflammatory meningeal conditions. The infections are not necessarily fatal; some of them are so slight as scarcely to claim close attention, more especially as they may be overshadowed by more urgent general symptoms, yet the congestion and effusion which they have induced may be sufficient to give rise to an occlusion of the route of the ventricular circulation, and subsequently to hydrocephalus.

FIG. 135.—Drawing from a specimen of ventricular hydrocephalus of the fourth degree. The condition developed as the result of a basal birth hæmorrhage. There is a hæmorrhagic effusion in the cisterna magna, which has occluded the foramina in the roof of the fourth ventricle.

An interesting example illustrative of this point is borne out by the following case-history :—

Case 1.—J. S. (6½ months). About six weeks before admission to hospital the baby had been much in contact with a number of cats who were suffering severely from distemper. The child took an illness which closely resembled the distemper of animals—temperature, coryza, discharge from nose and eyes. The child's uncle is an authority on veterinary matters, and he had no doubt that the child's illness was a direct infection from the animal source. Within ten days of the onset of the illness the child developed what clinically resembled a mild meningitis, with head retraction, vomiting, and eye squint. These symptoms lasted a few days, and a complete recovery then seemed to follow. About two weeks later it was noticed that the head was beginning to enlarge, and a hydrocephalus rapidly developed.

Investigation showed the hydrocephalus to be an obstructive one due to occlusion of the foramina in the roof of the fourth ventricle. So rapid was the enlargement of the head that operative interference was impossible. The child died, and subsequent examination demonstrated signs of a former basal meningitis, which had resulted in occlusion of the ventricular foramina.

FIG. 136.—Two examples of syphilitic hydrocephalus. Both cases were ventricular in type.

FIG. 137.—Drawing from a specimen of syphilitic basal meningitis. The condition has led to closure of the foramina of Magendie and Luechka, and to a resulting ventricular hydrocephalus of the fourth degree.

In certain cases of the series under consideration we believe that a former subacute basal meningitis was the essential causative factor.

d. Congenital Syphilis as a Cause.—In certain cases of congenital syphilis there may be an associated chronic inflammatory thickening of the cerebral meninges (Fig. 136). Such a change is most marked at the base of the brain (Fig. 137), and the more vascular pia-arachnoid is affected to a greater degree than the less vascular dura mater. It is in the tissues around the cisterna magna and the cisterna basalis that the plastic and adhesive specific meningitis may be most evident. We confess we have been surprised to find what a relatively large proportion of hydrocephalics are victims of congenital syphilis and show a positive Wassermann reaction. The meningeal changes may develop in the child who otherwise does not show the general stigmata

of the disease—in fact, it would appear that it is in the ill-defined and indefinite types of the disease that the meningeal changes are most in evidence.

It has further been our experience that in the atresic forms of hydrocephalus, where

the obstruction to the circulation of the cerebrospinal fluid is situated above the point of exit from the fourth ventricle, congenital syphilis is such a common concurrent condition that we believe that a considerable proportion of the atresiae are specific in origin. The point is one of considerable importance, because in certain cases in which the origin of the hydrocephalus was recognized as being specific, we were able to arrest the progress of the disease by means of antisiphilitic remedies without operative interference.

We believe that the possible origins of ventricular hydrocephalus may be summarized into three classes—according to whether it is dependent upon intracranial birth hæmorrhage, former subacute inflammatory meningeal condition, or the leptomeningitis which develops in association with congenital syphilis.

3. The Etiology of Extraventricular Hydrocephalus.—Until a recent publication of Dandy's work no satisfying explanation had been offered of the origin of the extraventricular type of hydrocephalus. It is clear that no obstruction exists up to and including the points of exit of the fluid from the fourth ventricle. The possibility that there is an over-production of cerebrospinal fluid has never been established, and, if this is not the fault, the only other possible explanation is a diminished absorption of the cerebrospinal fluid. The difficulty has been to explain the obstacle which exists to the absorption of the fluid.

Dandy's view as to the anatomical lesion responsible for this type has already been given, viz., pia-arachnoid adhesions which shut off the absorbing area of the cerebral sub-arachnoid space from the posterior cranial fossa.* In our series of cases, six have been examples of the extraventricular variety, and we have had an opportunity of satisfying ourselves that the explanation brought forward by Dandy has a great deal to recommend it. Certainly in these cases there is an extensive obliteration of the subarachnoid space by adhesions. They are best illustrated at autopsy, and, to demonstrate them efficiently, the brain should be removed with the dural covering, if possible, intact. From the base of the child's skull the dura can be removed with greater facility than is the case in the adult, and subsequent dissection will then demonstrate that the normal pia-arachnoid membrane is replaced in these cases by an opaque adherent membrane which extends over the region of the cisterna magna and basalis, around the cerebellar hemispheres, and downwards to the foramen magnum. Adhesions in this region must constitute a very real obstruction to the circulating cerebrospinal fluid, and, once the backward pressure has caused reaction and internal hydrocephalus has begun, a vicious circle is actually in existence, because, as the cerebral distention increases, the close contact of the cerebral tissues with the meninges and skull, and, more especially, with the unyielding base of the skull, further increases the obstruction to the upward circulation of the cerebrospinal fluid. The actual origin of the adhesions in extraventricular hydrocephalus is similar to that already discussed in relation to the ventricular type, with the proviso that birth hæmorrhages probably do not play a part in the first-mentioned variety: basal meningitis, simple or specific, is the more likely origin.

SOME POINTS IN THE MORBID ANATOMY OF HYDROCEPHALUS.

We have dealt with the more important points in the etiological pathology of the condition, and it remains to review the more outstanding characteristics of the morbid anatomy. The essential feature is a distention of the ventricular system with cerebrospinal fluid. The distribution of the distention will necessarily depend on the type of hydrocephalus. In the extraventricular type the complete ventricular system is involved; in the ventricular variety the distention will depend on the situation of the obstruction.

* Dandy states that he has produced communicating hydrocephalus experimentally by encircling the mid-brain with a strip of gauze saturated in iodine, and so inducing adhesions. *Ann. of Surg.*, 1919, lxx, 129.

The following table shows the distribution which has come under our notice.

Table 1.—THE TYPES OF HYDROCEPHALUS IN THE SERIES UNDER REVIEW.

Extraventricular type	..	6 cases	
Ventricular type, 1st degree	..	0	"
" " 2nd	"	0	"
" " 3rd	"	7	" (including 2 tumour cases)
" " 4th	"	8	"
Total	..	21 cases

FIG. 138.—A frozen section through the head of a ventricular hydrocephalus. It illustrates the extreme degree of destruction of the white matter. (*Dr. Thomson's case.*)

Our experience has been that the ventricular type is more common than the extra-ventricular type in a proportion of about 3 to 1, and of the ventricular type the fourth degree of obstruction, i.e., in the roof of the fourth ventricle, is the most common in a proportion of 8 cases out of 15.

The increasing distention of the ventricular cavities leads to a progressive thinning of the cerebral tissues, and it is interesting that the disappearance of the cerebral substance is at the expense of the white tissue—even in the most advanced example of the disease a zone of grey tissue of approximately normal thickness exists (*Fig. 138*). The cerebral

sulci become opened out, and sometimes they disappear ; this change is most marked in the frontal region of the brain. The fontanelles, the sutures, and the imperfect ossification generally of the cranial vault encourage an enlargement of the dome of the skull, while the more unyielding base is not correspondingly affected. It is this disproportion which gives the characteristic clinical appearance of hydrocephalus, and which also explains the unlikelihood of there being much displacement of the medulla downwards into the foramen magnum. We shall allude again to the importance of dealing with these cases before the destruction of cerebral tissues becomes marked ; the whole basis of successful treatment will depend on its early inauguration. As long as the basal ganglia remain intact life continues, and, as the distention follows the line of the yielding skull in an upward direction, these ganglia are wonderfully preserved from pressure until a late stage ; but, from the point of view of making operative interference worth while, close attention should be paid to the degree of destruction of cerebral white tissue which is proceeding.

A puzzling point in pathology is opened up when we attempt an explanation of a variety of hydrocephalus which is accompanied by a complete occlusion of the ventricular system, and yet the hydrocephalus has been spontaneously arrested. Through the kindness of a colleague we have had an opportunity of examining the case-records of two examples of this puzzling condition. The first and the more impressive case was that of a young adult, age 25 years, who had died suddenly from what was thought to be a cerebral hæmorrhage. There had been a complaint of giddiness and headache with some vomiting. Two hours later unconsciousness supervened, the breathing became stertorous, and within a short period he was dead. In the previous history there was no definite account of serious illness ; he had been recognized as a delicate man, nervous and highly-strung, constantly subject to headaches and attacks of migraine. The head was not unusually large, and on superficial examination there was nothing to suggest a hydrocephalus. On autopsy a peculiar state of affairs was discovered. The convolutions were flattened and relatively few in number, the entire ventricular system was dilated, and the roof of the fourth ventricle was completely occluded by a dense fibrous membrane. No trace could be discovered of the normal foramina, and tests applied showed that the intraventricular fluid had apparently no communication with the subarachnoid fluid along the usually recognized channels.

The second case occurred in a boy, age 8 years. We have no knowledge of the previous clinical history, but the autopsy specimen which we were permitted to examine showed a long-standing occlusion in the roof of the fourth ventricle, which, as far as we could discover, must have completely isolated the intraventricular from the extraventricular fluid.

The importance of cases such as these is that they would appear to indicate one of two possibilities :—

1. Either there are intraventricular media for the absorption of cerebrospinal fluid with which we are at present unacquainted ; or

2. Under certain conditions the isolation of the intraventricular fluid by occlusion of the foramina leads to a greatly diminished production of cerebrospinal fluid ; and further, it would seem to presuppose that there are extraventricular sources of production of cerebrospinal fluid, for in both of the above cases cerebrospinal fluid was present in the subarachnoid spaces.

It is clear, at any rate, that cases such as these indicate that there are possibilities in the physiology, or at least in the pathological physiology, of the cerebrospinal fluid circulation with which at present we are not completely acquainted.

There are many other details in the pathology of hydrocephalus to which we have not alluded ; we have purposely dealt only with those which have a bearing upon the surgical treatment of the disease.

CERTAIN CLINICAL FEATURES OF THE DISEASE.

Question of Sex and Age.—There is nothing to be learned from a consideration of the sex ; in the series under review there was a curious similarity in the sex occurrence, the figures being respectively 11 males and 10 females.

The age at which the cases came under surgical notice is shown in the following table :—

Table 2.—THE AGE OCCURRENCE.

	MONTHS												YEARS			
	1	2	3	4	5	6	7	8	9	10	11	12	2	3	4	5
Age ..																
Cases ..	—	2	3	5	1	1	2	1	2	—	—	1	1	2	—	—

The majority (18) of the cases were under one year old, 1 child was a year and a half, and 2 of the cases were between two years and three. No cases appeared at a later age than three years.

The Clinical History.—In certain cases the statement was offered that the parents noticed the head to be somewhat enlarged at birth, but it is doubtful if any real weight can be attached to this observation. It is clear that in no case was the head of such a size at birth as to offer any difficulty to delivery. There was no case, therefore, which would justify the qualification of being congenital in the sense that the signs of the disease were definitely present at birth. The two examples of true congenital hydrocephalus which have been mentioned are not included in this summary.

In certain instances it was remarked that the scalp veins were unusually prominent, and this feature was observed before any definite cranial enlargement was apparent.

As the head increases in size, the enlargement at first is a gradual one ; but after a definite point is reached the distention of the head progresses more rapidly. Dandy¹⁰ offers an observation which appears to contradict this statement, for he believes that the production of cerebrospinal fluid diminishes as the pressure within the ventricles increases ; but clinically we are convinced of the truth of the point which we have made. For example, such an observation as this was repeatedly made :—

E. H.—

First observation,	April 10,	circumference of head	17½ inches
Second	May 12	"	18 "
Third	June 15	"	18½ "
Fourth	July 10	"	21½ "
Final	July 30	"	23 "

During the last six weeks of observation the head increased by 4½ inches, as compared with an increase of one inch during the first two months of observation. It would seem that, after a definite point is reached, the tension exerted by the skull is so reduced that the underlying cerebral distention is less restricted than before. There is no clinical evidence that there is a diminished production of cerebrospinal fluid as the tension increases.

As the head enlarges the axis of the eyes is displaced downward, so that the sclerotics appear constantly underneath the upper lids. There is often a well-marked strabismus. The occurrence of nystagmus is an unusual feature. In the later stages of the disease sluggishness of the pupils and atrophy of the optic nerve are sometimes present. The mentality of the children ranges between extreme idiocy and normal intelligence. Most of the patients are feeble-minded and apathetic ; if old enough, they have difficulty in speaking. Motor disturbances are usually well-marked, and manifest themselves in spasms, paresis, unusual rigidity, and tremors ; the lower extremities are more severely affected than the upper. Sometimes the spasms and paresis are more marked on one side than on the other. The reflexes are increased. Twitching of individual groups of muscles and general convulsions may occur. Hydrocephalic children usually present a delicate appearance, they are pale and emaciated, with a senile expression. On account of the weight of the head its support is difficult, and therefore it is often bent forwards or thrown backwards. These children generally cry a good deal, and they do not readily put on weight. The appetite is not bad, but digestion is usually retarded.

It is sometimes remarkable how few general symptoms may exist in a hydrocephalus of the most marked degree. The infant a section through whose head is illustrated in *Fig. 138* was one of this type. It can be recognized how very marked was the atrophy of nerve-tissue, and yet, to within forty-eight hours of death, the intelligence was such that the child recognized its mother, while there was very little evidence of paralysis or rigidity of the superficial musculature.

THE PHYSICAL EXAMINATION OF THE CASE.

In the majority of cases it is sufficiently obvious that a hydrocephalus is present, but further examination is required if the case under review is to be efficiently classified; and efficient classification necessarily involves that an attempt be made to answer three questions: 1. *Does any evidence exist which would indicate the possible origin of the disease?* 2. *To what variety of hydrocephalus does it belong? Is it ventricular or extraventricular in type?* 3. *If it is ventricular in type, at what level does the lesion exist which is responsible for the hydrocephalus?*

The physical examination is directed towards supplying an answer to each of these individual queries so far as is possible. The answers having been given, classification is possible, and an intelligent treatment may be planned.

1. **The Origin of the Disease.**—If this question can be answered, the information will be obtained by careful questioning of the parents, a full case-history, and general examination of the child. Attention is specially paid to three possibilities—the occurrence of a syphilitic infection, the infliction of an injury to the skull, such as may have produced an intracranial hæmorrhage, and the history of a previous meningeal or encephalic infection. A fourth possibility exists, namely, the possibility of the coincident existence of a cerebral tumour.

In certain cases the evidence obtained is sufficiently strong to enable one to estimate with a degree of certainty the origin of the hydrocephalus, as, for example, in the specific types and the post-infective meningeal conditions.

A considerable proportion of cases, however, necessarily remain in which no definite idea can be formed of the etiology, and such a lack of knowledge is not after all serious, because an answer to the question is not essential in deciding on the line of treatment to be adopted. If possible, however, an attempt should be made to classify the case according to its etiology.

2. **The Type of the Hydrocephalus.**—According to Dandy's classification the question would be put: Is the hydrocephalus obstructive or communicating in type? But we have explained our reasons for preferring to express the varieties as ventricular or extraventricular.

Recognizing that practically all cases of hydrocephalus are obstructive in type, it is necessary to demonstrate whether the obstruction exists somewhere in the ventricular system up to and including the point of exit of the fluid from the fourth ventricle, or whether it exists in the subarachnoid spaces (communicating). Three procedures are followed in demonstrating the answer to the question:—

a. **Lumbar Puncture.**—Lumbar puncture may give a suggestive result, but not a definite answer to the question. An intraventricular hydrocephalus may show low tension in the spinal fluid, while an extraventricular hydrocephalus may show an increased tension; but variations exist, and occasionally the sequelæ are reversed. Therefore, while this investigation may be suggestive, it cannot be accepted as absolute.

b. **Intraventricular Injection of an Indicator, with Investigation of the Cerebrospinal Fluid.**—Following Dandy's recommendation, we have used phenolsulphonaphthalein as an indicator. It is important that the solution employed should be neutral, and we have employed the preparation supplied by Martindale. Inattention to the detail of the neutrality of the fluid may result in a sharp reaction characterized by temperature and signs of cerebral irritability.

The technique of injection is simple: one or other lateral ventricle is punctured with

the needle of a 'record' syringe, the syringe holding 1 c.c. of the phenolsulphonephthalein solution. When the ventricle has been entered 1 or 2 c.c. of cerebrospinal fluid are removed into the syringe barrel and allowed to mix with the indicator. The mixture is then injected into the ventricle, and the needle withdrawn. After an interval of time lumbar puncture is done, and the spinal fluid is allowed to pass into a test-tube containing a few drops of 25 per cent sodium hydrate solution.

Dandy recommends that thirty minutes should be allowed to elapse before the lumbar puncture is done; in our experience such a long interval is unnecessary. If the indicator can be recovered in the spinal fluid it will become obvious within five minutes, and sometimes even less.

By the recovery of the indicator (as evidenced by the pink coloration of the cerebrospinal fluid when it comes into contact with the sodium hydrate) it may be accepted that the hydrocephalus which exists is not due to an intraventricular obstruction up to and including the point of exit of the fluid from the fourth ventricle. In other words, the recovery of the indicator means an extraventricular hydrocephalus (communicating type—Dandy), while the non-appearance of the indicator in the spinal fluid may be taken as demonstrating an intraventricular hydrocephalus (obstructive type—Dandy).

c. The Recovery of the Ventricular Injection Substance from the Urine.—It is said that normally the amount of fluid absorption which occurs in the ventricular system is negligible; it is estimated at less than 1 per cent in two hours. The bulk of absorption within the skull occurs in the subarachnoid spaces, from which area as much as 40 to 60 per cent of an introduced fluid is excreted after two hours' interval. Based on these considerations the recovery of phenolsulphonephthalein from the urine subsequent to its injection into the ventricles becomes a matter of importance, for if a ventricular hydrocephalus exists practically none of the indicator will be recoverable within a reasonable time (two hours), while if an extraventricular hydrocephalus is present, the indicator will be recoverable, though not to the same degree as in a normal case.

We have not put this method of investigation into routine use. It expresses the same knowledge as is gained from an examination of the spinal fluid, while its demonstration is a matter of greater difficulty. Therefore we have relied on answering the question regarding the type of hydrocephalus by the second method of investigation, the examination of the spinal fluid after the intraventricular injection of an indicator.

Investigation by these methods, and more especially by the second, carries the observer a step forward, in so far as he is now able to classify the case of hydrocephalus into a ventricular or an extraventricular type. If it should happen that the case is an extraventricular one, we have not considered it necessary to proceed further with the routine examination.

By the injection of air into the spinal theca Dandy¹¹ has demonstrated after x-ray examination the actual location of the adhesions which are responsible for this type of hydrocephalus. He has shown the arrest of the air at the base of the brain and its absence from the cerebral sulci. All these points are demonstrable in extraventricular hydrocephalus, but their demonstration does not aid us in the question of treatment, at least, with the methods at present available to us.

3. The Level of the Lesion.—If, however, our original investigation has shown that the hydrocephalus is a ventricular one, further details of knowledge must be available before an intelligent operative treatment can be undertaken. The degree of hydrocephalus must be known, the situation of the obstruction must be localized: this knowledge is gained by the method of ventriculography.

The Technique of Ventriculography.—Cerebral diagnosis and cerebral surgery would benefit if it were possible to introduce into the ventricular system a fluid opaque to the action of x rays. Hitherto no fluid has been suggested which fulfils the necessity of opacity and yet does not irritate the delicate structure of the ependyma and choroid plexuses. The necessity of the non-irritating character of the fluid is all the more important when we consider that in intraventricular hydrocephalus the fluid may be isolated within the ventricles for a prolonged period of time—in fact, until it is artificially liberated or removed.

In the absence of a safe and yet efficient fluid, air has been employed, and in young children, in whom the ossification of the skull is not too advanced, it delineates with remarkable distinctness the ventricular outlines. The method is simple. One lateral ventricle is punctured, and to the needle an accurately fitting 'record' syringe of 20-c.c. capacity

FIG. 139.—The position of the head for ventriculography. A, To record the outline of the left lateral ventricle. B, To record the outlines of the third and fourth ventricles.

is fitted. The cerebrospinal fluid is slowly removed from the cavity of the ventricle, and when 20 c.c. are evacuated, a corresponding amount of air (20 c.c.) is injected into the ventricle, using the empty 'record' syringe for this purpose. The joint between needle and syringe is covered with sterile vaseline to prevent an escape of air at this point.

In a hydrocephalus of moderate size (18 to 19 inches in circumference) we remove 70 to 80 c.c. of fluid, replacing the fluid with a corresponding quantity of air. The removal of the fluid and the introduction of the air must be carried out in small alternating amounts, as the sudden removal of a large quantity of fluid without the supporting influence of the air might induce a cerebral cedema, and possibly an intraventricular hæmorrhage. After the introduction of the air, a hollow note can be elicited over the ventricle, and on movements of the head air may be heard to gurgle from one portion of the ventricular system into another.

FIG. 140.—Ventriculogram showing outline of dilated lateral ventricle filled with air. (Lateral position of head.)

Radiograms are now taken with the head in three different positions—right and left at lateral, and with the head hanging downwards so that the skull base is at the highest level (Fig. 139). The first plate will show the outlines of one lateral ventricle (Fig. 140), the

second will demonstrate the opposite ventricle—assuming that the foramina of Monro and the third ventricle are patent—while the third plate will illustrate the third and fourth ventricles and the iter.

FIG. 141.—Topography of normal lateral ventricle.

Such views of the ventricular system would probably be very difficult to demonstrate in the normal brain (*Fig. 141*). We have never made the attempt, but in the hydrocephalic, where the ventricular system is distended, definite and clear representations can be obtained. The value of the examination lies in the fact that it will locate the level at which the obstruction exists.

We have not had an opportunity of observing the appearance of an obstruction in the foramen of Monro; iter obstructions are characteristic (*Fig. 142*); obstruction in the roof of the fourth ventricle is also distinctive (*Fig. 143*), for, though ventriculography of an extraventricular hydrocephalus gives a somewhat similar representation, it can be otherwise excluded as shown above.

The introduction of air into the ventricles may appear to be a somewhat heroic proceeding in the investigation of a case of hydrocephalus,

but actually the method is practically devoid of risk; certainly with the cases which

FIG. 142.—Ventriculogram showing outlines of dilated left lateral and third ventricles. The air has failed to enter the aqueduct of Sylvius, indicating an obstruction at this site—ventricular hydrocephalus of the third degree. (Inverted position of head.)

FIG. 143.—Ventriculogram showing outlines of dilated left lateral, third, and fourth ventricles. Note the air in the right foramen of Monro, and the bulging roof of the fourth ventricle. Obstruction at the roof of the fourth ventricle is indicated. Ventriculogram in conjunction with the coloured indicator test. (Inverted position of head.)

have come under our care we have had no anxiety. Occasionally there is a reactive rise of temperature, but it is not excessive, and it soon subsides:

A Survey of the Clinical Examination.—A successful clinical examination will have thrown some light on three aspects of the disease : (1) The possible origin ; (2) The type—ventricular or extraventricular ; (3) The degree of the ventricular type. With these facts at our disposal the consideration of the treatment may reasonably be discussed.

TREATMENT.

The Importance of Early Treatment.—If the efficient treatment of hydrocephalus is to give us satisfactory results it must be carried out before the expansion of the head has become too marked. There is nothing to be gained from operating on a child if the white matter of the brain is already reduced to a shell. No definite limit can be fixed ; it must be left a good deal to the decision of personal judgement ; but we have found that an increase in circumference measurement up to 3 inches above the normal is within the range of satisfactory post-operative recovery. Above this figure we enter the range of unsatisfactory results, and with every degree of increase the probability of improvement diminishes.

Various Lines of Treatment hitherto Suggested.—Kausch,¹² in the course of a most exhaustive article, details the various procedures which have hitherto been tried in the treatment of hydrocephalus. We append the following tabular account ; there is nothing to be gained by a more detailed recitation of the methods, and those who are interested are referred to Kausch's contribution (*see also* Haynes¹³).

The following methods have been suggested or actually used in the treatment of hydrocephalus :—

1. Intermittent drainage	{	i. Of lateral ventricle
		ii. Of the spinal canal
2. Continuous drainage	{	iii. By puncture of the corpus callosum
		i. To the surface
		ii. To subcutaneous tissue
A. Of the lateral ventricle	{	iii. Into subdural or subarachnoid spaces
		iv. Into the peritoneal cavity
		v. Into the temporal vein
		vi. Into the superior longitudinal sinus
B. Of the spinal canal	{	i. To surface
		ii. Into retroperitoneal tissues
		iii. Into peritoneal cavity
C. Of the subarachnoid space (fourth ventricle)		
D. Of the cisterna magna into the cranial sinuses		
3. Indirect treatment (carotid ligatures)		
4. Other methods of treatment, such as by a seton, injection of iodine, galvanopuncture, compression of head, and by drugs		

With such a variety of methods of treatment available, the conclusion may be come to that no single one has proved efficacious. Incomplete methods of investigation and an imperfect knowledge of the etiological pathology have been responsible for this unsatisfactory state of affairs.

The Essential Difference in Treatment between the Ventricular and the Extraventricular Types.—It is obvious that a very clear distinction must be drawn from the operative point of view between the ventricular and extraventricular varieties. Both are certainly obstructive in nature ; but, while one (ventricular) is localized and accessible, the other (extraventricular) is diffuse, and practically inaccessible.

If the hydrocephalus is of the ventricular variety—that is to say, if there is an obstruction at some portion of the ventricular system proximal to and including the roof of the fourth ventricle—experience has shown that the only efficient way of treating the hydrocephalus is by removing the obstruction, and so opening up the normal passage for the circulation of the cerebrospinal fluid.

In the extraventricular variety the problem is more difficult ; but we discuss later the possibilities of treatment of this variety.

THE OPERATIVE TREATMENT OF VENTRICULAR HYDROCEPHALUS.

All examples of ventricular hydrocephalus, with the exception of the very rare first and second degrees, are operated on by the occipital route. We have not had an opportunity of treating a case of the first or second degree, and would therefore refer the reader to Dandy's observations on this point. The essential feature of the suboccipital operation is to expose that portion of the ventricular system which is most accessible, namely, the roof of the fourth ventricle, and through this region to deal with or remove the obstruction which exists.

FIG. 144.—*The suboccipital operation for hydrocephalus: 1.*—The position of the head upon the head-rest is shown. The incision is indicated by the dotted line. The self-retaining retractor has been fixed in place by a rubber band round the head. The anæsthetic is being administered by the intrapharyngeal method.

Preliminaries and Anæsthesia.

—The operation should be undertaken while the child is in as fit a condition as possible. Nothing should be attempted if there is any suspicion of cold or bronchitis. Throughout the operation, which may be a lengthy one, the child is kept warm—an arrangement like a sleeping bag made of gamgee tissue meets the requirement. The child is placed on its face with the head flexed on the chest and, to ensure a satisfactory position, we have adopted a special head-rest which supports the head in the correct position and permits the administration of the anæsthetic and yet does not interfere with the respiratory movements (*Fig. 144*).

Intrapharyngeal ether has been the anæsthetic of choice, the anæsthetic being administered by a catheter introduced through the nostril.

The Exposure of the Roof of the Fourth Ventricle.—A mid-line incision is made, extending from the external occipital protuberance downwards to the level of the 7th cervical spine. In the earlier cases we employed a crossbow incision, but it involved an unnecessary amount of hæmorrhage, and the access which it provides is not appreciably greater than

FIG. 145.—*The suboccipital operation: 2.*—The vertical muscles of the suboccipital region have been split in the middle line, down to the bone. The periosteum has been detached from the occipital bone. These structures are retracted. The spine of the axis, with the attachments of the deep muscles, is exposed. The bone has been perforated with the burr over either cerebellar hemisphere.

that afforded by the straight mid-line incision. The incision is deepened through the fibrous intermuscular space, exposing the mid-line of the occipital bone and the spines of the upper cervical vertebræ (*Fig. 145*). The muscular attachments are separated outwards, and such free separation is possible that we have not found it necessary to carry out any transverse division of the muscles. We believe that any transverse section of the muscles is to be avoided; the upper portions of the muscles in the child are flimsy, and they are easily so bruised and destroyed as to make their re-attachment difficult. From the occipital bone the separation may be carried out subperiosteally; the trapezius need not be disturbed, but the complexus, the rectus capitis posticus minor, the superior oblique, and the rectus capitis posticus major are separated outwards to the line of the occipital artery; this vessel is not in view, as it is under cover of the separated muscles. The result is an exposure of the occipital bone, vertically from the 'linea suprema' to the margin of the foramen magnum, and laterally from the right to the left occipital artery line.

Two trephine openings are now made, one on each side of the mid-line, so as to avoid the occipital sinus. The trephine openings are made with a Hudson's drill, and enlarged with a burr. With a rongeur forceps a crescentic area of bone is removed, the concavity of the crescent being at the posterior margin of the foramen magnum, while the convexity is half an inch within the area of the muscular separation. The removal of the posterior margin of the foramen magnum is an important point. During this stage of the proceedings bleeding is troublesome, more especially at either side of the foramen magnum; it is arrested by the use of bone wax and by gauze plugs. Special care is required as the area of bone over the occipital sinus is removed, and the downward removal of bone should not be carried so far as to damage the emissary veins which pierce the bone a quarter of an inch from the lateral angles of the foramen magnum. The upward removal of bone need not expose the lateral sinuses.

The dura of the cerebellar fossa appears in view, and an estimation can be formed of the degree of intracranial pressure which exists by the amount of dural bulging which is present. If there is considerable tension we now puncture one lateral ventricle and remove a sufficiency of fluid to relieve appreciably the pressure which exists.

The next stage is the control of hæmorrhage from the occipital sinus and from the marginal sinuses. This is efficiently done by including the various sinuses in sutures carried on round needles. Four ligatures are applied, one at each extremity of the occipital sinus and one around each marginal sinus as far forward as the ligature can conveniently be placed (*Fig. 146*).

The dura is now opened over each cerebellar hemisphere, and it is carefully separated inwards until the falx cerebelli is reached. This structure is divided about its centre with a fine pair of blunt-pointed scissors, and immediately there is a retraction of the divided dural tissue and an exposure of the posterior surface of the cerebellar hemisphere covered by the pia-arachnoid tissues. Additional space can be obtained, when necessary, by

FIG. 146.—*The suboccipital operation*: 3.—The bone defect has been completed and the posterior margin of the foramen magnum removed. Ligatures have been applied to control the occipital and marginal sinuses. The dura mater has been incised on either side of the mid-line.

extending short vertical incisions upwards from either extremity of the transverse incision, so as to form an upper dural flap. A special retractor, having a cup-shaped surface for

FIG. 147.—*The suboccipital operation: 4.*—The dura mater has been completely opened, and the upper flap allowed to retract. The cerebellar hemispheres and cisterna magna are exposed.

each cerebellar hemisphere and a central notch which is occupied by a small electric lamp, is now introduced beneath the cerebellum, and that organ is gently displaced upwards. The displacement must of course be regulated with the greatest delicacy and care. A space is exposed which lies between the under surface of the cerebellum and the posterior surface of the medulla, and which is bridged across by strands of arachnoid tissue; this is the cisterna magna (*Fig. 147*).

If the hydrocephalus is of the fourth degree, the obstruction having occurred in the roof of the fourth ventricle, and if it is of an adhesive type, the result of a birth hæmorrhage or a former basal meningitis, this space may be largely obliterated by fibrous adhesions and hæmorrhagic extravasations. If such is the case, great care must be exercised in the separation of the adhesions, because, owing to the distention of the ventricles, the roof of the fourth ventricle is displaced backwards, and is torn before one is aware of its proximity. The roof of the ventricle should now come into view. If it is a bulging tent-like structure with definitely thickened texture and obvious obliteration of its foramina, it may be accepted that it is the site of the obstruction of the circulation of the cerebrospinal fluid, and be dealt with accordingly. It is our practice in such cases to remove a diamond-shaped area of the tela choroidea with fine spring forceps and eye scissors fitted with long-angled handles (*Fig. 148*).

The Relief of an Iter Obstruction.—If it should happen that the hydrocephalus is of the third degree, the obstruction existing in the iter, the operation has to be carried a stage further. In the third degree the fourth ventricle is not distended, and the obstruction in the iter is generally at one or other extremity. To gain access to the iter we follow the technique which Dandy first recommended. A small nasal speculum is inserted beneath the inferior vermis of the cerebellum, and, if further access is necessary, the vermis is split in the mid-line with a fine-bladed knife. The edge of the inferior medullary velum comes into view, and,

the cerebellar retractor has been introduced and the cerebellum displaced upwards and its lobes laterally. The roof of the fourth ventricle is in view, illuminated by the small lamp. The foramina of Magendie and Luschka are seen (if not obliterated by adhesions).

FIG. 148.—*The suboccipital operation: 5.*—The cerebellar retractor has been introduced and the cerebellum displaced upwards and its lobes laterally. The roof of the fourth ventricle is in view, illuminated by the small lamp. The foramina of Magendie and Luschka are seen (if not obliterated by adhesions).

The edge of the inferior medullary velum comes into view, and,

if the tela choroidea forming the lower half of the ventricular roof has not been opened up, it is now perforated. A fine rubber catheter is guided into the upper angle of the ventricle, and is gently pushed onwards. The obstruction may be of such a fragile nature that there is little real resistance, and the catheter passes into the dilated third ventricle. This is signaled by a gush of cerebrospinal fluid. If the obstruction is more definite and cannot be overcome by the catheter, a millimetre probe is used to open up the closed communication (*Fig. 149*); this instrument, however, must be used with extreme care, as any forcible manoeuvre may very easily result in perforation of the brain tissue around.

After the channel has been opened with the probe the small rubber catheter is inserted, and it is demonstrated by the free escape of cerebrospinal fluid that the communication has been established.

As regards the further procedure, Dandy recommends that the rubber catheter should be left *in situ* for several weeks, the lower end, cut short, being coiled up in the cisterna magna. He believes that this permits the formation of an epithelial lining around the tube, and diminishes the possibility of secondary closure. At first we followed this advice, but in the later series of cases we have been

satisfied with efficient canalization of the iter, and it would seem that the restoration of the flow of the cerebrospinal fluid is sufficient to keep the channel open.

The Closure of the Wound.—If the operation is to be a successful one, the relief of the obstruction must be followed by the most careful closure of the wound. A leak of cerebrospinal fluid to the surface is unlikely to become spontaneously arrested, and it ends almost certainly in sepsis and death.

An attempt should be made to bring the dural edges together with a continuous catgut suture; the relief of tension which has resulted generally makes this procedure possible. The muscles are then closed in two layers, and the advantage of the straight incision over the crossbow incision now becomes evident, because the closure can be accomplished so much more efficiently.

In the immediate post-operative period attention must be paid in case the intracranial tension increases to such an extent as to lead to a leakage of fluid. If the tension is becoming marked, ventricular puncture should be practised, and if necessary repeated.

THE TREATMENT OF EXTRAVENTRICULAR HYDROCEPHALUS.

In this type of the disease the problem is in some respects more difficult than in the case of the ventricular variety. The obstruction is a widespread adhesive one at the base of the brain, and it is therefore inaccessible to direct removal. It has to be borne in mind, however, that certain cases of the extraventricular variety show a spontaneous arrest of the disease, a possibility which can never be hoped for in the intraventricular variety.

At the present time the attempts at treatment have been directed towards diminishing

FIG. 149.—*The suboccipital operation: C.*—Final step to relieve an obstruction in the aqueduct. The roof of the fourth ventricle has been opened and the lower part of the vermis split. With the aid of the nasal speculum the upper terminus of the fourth ventricle is exposed. A graduated probe is inserted through the aqueduct into the third ventricle.

the production of the cerebrospinal fluid to such an extent as to bring it within the power of absorption which exists, for of course a certain degree of absorption still continues. There are two methods which have been used in securing this diminished production of cerebrospinal fluid.

Dandy¹⁴ has advised the removal of the choroid plexus from within the lateral ventricle, and he has published successful results in several cases. We have hitherto been content with less drastic measures. Intraventricular plexectomy is an exceedingly grave operation to perform, and there is no physiological proof that removal of a limited amount of choroid plexus proportionately diminishes the production of the cerebrospinal fluid. Stiles suggested the possibility of diminishing the production of the fluid by ligation of the common carotids, and in 1898¹⁵ and again in 1912¹⁶ he reported favourable results from this operation. It is clear, of course, that it is only the extraventricular type which will benefit from this procedure; its employment in the ventricular variety will inevitably lead to disappointment, and this may to some extent explain the adverse criticism which the method has received.

We have treated all the examples of extraventricular hydrocephalus which have come under our care by ligation of the common carotids, and the results have been sufficiently promising to warrant the continuation of this method. The actual results are

communicated later. The technique of the operation requires no detailed description—the vessels are tied at the crossing of the omohyoid muscle at an interval of ten days.

We confess that it seems unreal that ligation of the common carotid vessels should result in a diminished production of cerebrospinal fluid, as, through the medium of the anterior choroidal vessels, they are responsible for only a portion of the blood-supply of the choroid plexus. Nevertheless, we have definite clinical evidence that, following the procedure of ligation, a certain proportion of cases of extraventricular hydrocephalus become arrested. The procedure is such a simple one that it seems worth a more extended trial in the special type of case for which it is suited.

But the methods of plexectomy and carotid ligation are at best unsatisfactory. No definite result can be guaranteed, because they do not deal with the pathological condition which is responsible for the hydrocephalus, the subarachnoid ob-

FIG. 150.—Suggested operation for the relief of certain types of extraventricular hydrocephalus. The pathway of the cerebrospinal fluid to the absorbing area of the cerebral subarachnoid space has been obstructed by adhesions between the free edge of the tentorium and the adjacent brain. By entering the skull above the lateral sinus and elevating the occipital lobe an opening has been cut in the tentorium outside the adherent area. The arrow indicates the new pathway for the cerebrospinal fluid.

struction. Considering the problem from the etiological point of view, the obvious remedy is to open up a new pathway for the fluid in order to permit it to pass over the absorption area of the cerebral lobes. If the adhesions exist at the base of the brain, occluding the cisterns, it is difficult to see how any direct relief can be carried out; but if the adhesions occur between the free edge of the tentorium and the mid-brain, there are greater possibilities of operative interference. In the cadaver we have been able to carry out a procedure which we hope after further trial to employ in a certain variety of extraventricular hydrocephalus, the variety which is shown by ventriculography to be the result of adhesions between the mid-brain and the free edge of the tentorium. The proposed procedure is illustrated diagrammatically in the accompanying sketch (Fig. 150). The skull is trephined immediately above the lateral sinus, midway between the

mastoid process and the external occipital protuberance. The dura is opened, and the occipital lobe is elevated; the upper surface of the tentorium is exposed, and this is carefully divided radially without damage to the lateral sinus: the incision through the tentorium is prolonged inwards as far as is safely possible. Through the incision thus made it is conceivable that a fresh channel may be established for the circulation of the fluid. The procedure is still hypothetical as far as its practical value is concerned, but it offers possibilities which we hope to test.

RESULTS.

Tables 3 and 4 summarize the cases and the results obtained. Two cases of hydrocephalus associated with tumours are not included in this summary.

Table 3.—VENTRICULAR TYPE.

NO.	NAME AND AGE	DEGREE	RESULT
1	J. S., 6½ months	4th	Died
2	E. A., 4½ months	3rd	Died
3	E. H., 9 weeks	4th	No operation
4	A. Y., 10 months	4th	Died
5	M. D., 5 months	3rd	No operation
6	R. M., 3 months	4th	Died
7	C. F., 7 months	4th	Cure
8	D. T., 7 months	4th (specific type)	Improved
9	N. S., 3 months	4th	No operation (died)
10	T. C., 1 year 4 months	3rd	Died
11	M. M., 4 months	4th	Arrested (probable cure)
12	M. C., 3 years	3rd	Improved
13	J. H., 6 months	3rd	Cure

Analysis of the Results of the Ventricular Type.—At first sight the results appear to be exceedingly unpromising—out of 13 cases, in 3 the condition was so advanced as to preclude any prospect of success from operative interference, 5 cases succumbed from the operation, in 3 cases the operative interference has resulted in apparent arrest of the disease, but we do not classify these as cures, because there has been no diminution in the bulk of the head, and the mental condition has remained permanently impaired; it is unlikely that these children will ever become useful members of society, capable of taking their share in the work of the world. In 2 instances (and this is the hopeful side of the problem) it would appear that we have been successful in effecting a complete cure. The following is a brief epitome of the histories of these cases:—

Case 1.—C. F., 7 months. Female. Recommended by Dr. Brander, Fort William. On admission to hospital the child (*Fig. 151*) was suffering from a marked hydrocephalus; the fronto-occipital circumference was 20½ inches. The size and weight of the head was so great that it continually fell forward on the chest. Investigation showed that the hydrocephalus was ventricular, and of the fourth degree. The child was operated on in May, 1921, and, as a matter of fact, it was one of the cases done during the afternoon demonstration at the Children's Hospital when the British Association of Surgeons met in Edinburgh. An obstruction was found in the roof of the fourth ventricle, and this was relieved. An excellent post-operative recovery was made.

FIG. 151.—Case of hydrocephalus, C. F. Appearance of child before operation. The hydrocephalus was a ventricular one of the fourth degree.

One year has now elapsed since the operation, and the accompanying photograph (*Fig. 152*) bears witness to the child's present condition. The head has shrunk in size, the fontanelles are closed, and the sutures well ossified; the general intelligence of the child is unimpaired; it stands without assistance, and it is beginning to walk. The only evidence of defect which remains is an internal strabismus present in both eyes (*Fig. 153*).

FIG. 152.—Case of hydrocephalus, C. F. Appearance of child one year after operation. An obstruction in the roof of the fourth ventricle was removed.

FIG. 153.—C. F. Operated on for a ventricular hydrocephalus of the fourth degree when 7 months old. Present appearance of the child.

Case 2.—J. H., 6 months. Male (Dr. Ainslie, Edinburgh). The child, on admission to hospital, was found to be the victim of a ventricular hydrocephalus of the third degree. It was operated on in October, 1921, and the obstruction in the iter was relieved. A good recovery was made.

Nearly seven months have now elapsed since the operation was performed, and a complete recovery appears to have been made. The head has diminished in size, and ossification is apparently complete except at the anterior fontanelle. The child is a sharp intelligent baby, and present appearances indicate normal development (*Fig. 154*).

FIG. 154.—J. H., age 12 months. Seven months after operation for a ventricular hydrocephalus due to an iter destruction.

This proportion of cases (2 out of 13) is, of course, exceedingly small, but there are indications which promise better in the future. The operative procedure is of a highly technical character, and considerable experience is necessary if certain errors are to be avoided. Evidence of this is brought out by the fact that of the last 7 cases only 1 has succumbed after operation, 2 have been cured, 3 have been improved in so far as the condition has apparently been arrested, while one case was so extensive as to permit of no operative interference. We hope that in the future the knowledge we have gained may still further improve the results.

An Analysis of the Results of the Extraventricular Type.—In this group there were 6 cases: only 5 were submitted to operative interference (ligature of carotids), one succumbing on admission to hospital. Of the remaining 5 cases, 1 succumbed within forty-eight hours of the first operation, as the result of hyperpyrexia, and 4 were successfully operated on. Of these, 3 have very definitely improved, and we believe that the hydrocephalus is now

arrested ; in the fourth case no improvement followed the operation, and the hydrocephalus increased to a fatal issue.

Table 4.—EXTRAVENTRICULAR TYPE.

NAME AND AGE	METHOD ADOPTED	RESULT
R. W., 4 months	Ligature of carotids	Condition arrested
S. S., 8½ months	" "	Condition arrested
M. M., 1 year 2 months	" "	Condition progressed (fatal in six months)
E. S., 9 months	No operation	Succumbed on admission
N. M., 9 months	Ligature of carotids	Condition arrested
A. P., 10 months	Ligature of one carotid	Succumbed to hyperpyrexia within 24 hours of operation

Out of a total of 4 cases of extraventricular hydrocephalus with carotid ligature, 3 have very definitely benefited (*Figs. 155 and 156*), and, though we do not wish at this stage to classify them as complete cures, we believe that everything points to a successful issue. The total series is, of course, too small to afford conclusive evidence, but the proportion of 3 improvements out of 4 cases has so encouraged us that we intend to continue to practise this method of carotid ligature in cases of extraventricular (communicating) hydrocephalus in preference to the method of plexectomy.

FIG. 155.—S. S., age 8 months. An example of an extraventricular hydrocephalus. Before operation.

FIG. 156.—S. S. Extraventricular hydrocephalus. The same child as shown in *Fig. 155*, six months after ligature of carotids.

REFERENCES.

- ¹HILTON, *Rest and Pain*, 1876, 38.
- ²DANDY, W. E., and BLACKFAN, K. D., *Jour. Amer. Med. Assoc.*, 1913, lxi, 2216; *Amer. Jour. Dis. Child.*, 1914, viii, 406; *Beit. z. klin. Chir.*, 1914, xciii, 392.
- ³DANDY, W. E., *Ann. of Surg.*, 1919, lxx, 129.
- ⁴SPINA, A., *Arch. f. d. g. Physiol.*, 1899, Bd. 76, 420.
- ⁵WEED, L. H., *Jour. of Med. Research*, 1914, xxxi, 21.
- ⁶WEED, L. H., *Ibid.*, 51.
- ⁷DANDY, W. E., *Ann. of Surg.*, 1918, lxxviii, 570.
- ⁸DANDY, W. E., *Ibid.*, 1919, lxx, 134.
- ⁹DANDY, W. E., *Amer. Jour. Dis. Child.*, 1917, xiv, 424.
- ¹⁰DANDY, W. E., *Ann. of Surg.*, 1918, lxxviii, 576.
- ¹¹DANDY, W. E., *Ibid.*, 1919, lxx, 397.
- ¹²KAUSCH, *Arch. f. klin. Chir.* 1908, Bd. 87.
- ¹³HAYNES, I. S., *Ann. of Surg.*, 1913, lvii, 449.
- ¹⁴DANDY, W. E., *Ann. of Surg.*, 1918, lxxviii, 569.
- ¹⁵STILES, *Brit. Med. Jour.*, 1898, ii, 1157.
- ¹⁶STILES, *Surg. Gynecol. and Obst.*, 1912.