Cases of Excessive Hydrocephalus with Ruptured Diverticulum of the Third Ventricle, Caused by Aqueductal Malformation

By

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According to Dandy [5] 95 per cent, according to Russel [17] 99 per cent of all the cases of hydrocephalus are due to an obstacle in the circulation of the cerebospinal fluid (mechanical, occlusive hydrocephalus) and it is only in the remaining 5 per cent and 1 per cent, respectively, that overproduction or disturbance in the absorption of the CSF has any role to play (functional hydrocephalus).

In the case of mechanical hydrocephalus the causes of occlusion may be

(i) Developmental malformation [2, 3, 8, 9, 12, 13, 14, 15, 16]

(ii) Inflammation [5]

(iii) Tumour [1, 2, 9]

(iv) Haemorrhage [17]

In the following we shall deal only with the problem of developmental disorders. In such cases the occlusion is found most often in the region of the aqueduct of Sylvius.

The developmental disorder of the aqueduct may be a stenosis or an atresia. In the case of stenosis we find an aqueduct normal in histological structure, but narrower than usual in diameter. The latter is difficult to estimate, because the aqueduct's diameter is highly variable even under normal conditions [2, 4, 6]. The traction stenosis of the aqueduct may be a complication of Arnold-Chiari's complex. Complete atresia of the aqueduct has been described by many authors, but according to Russel [17] there is no area in the histological sections of such cases in which the lumen would have disappeared completely, thus, according to that author the term atresia is incorrect from the pathological point of view, the proper term being "forking". There are namely several, rudimentarily developed aqueducts in such cases, that either terminate blindly, or communicate with each other or the fourth ventricle. In these narrow rudimentary canals CSF flow is very slight or even impossible. A less common developmental disorder of the aqueduct is the septum formation, which obstructs the aqueduct normally developed in other areas [14, 17, 21].

Case Reports

Case 1.

a) *Clinical Data*

N. Zs., a female baby born from the first pregnancy. During the seventh month

of pregnancy the mother had been under observation with the suspicion of appendicitis. The infant had been born prematurely, with a weight of 2090 g and a skull circumference of 30.5 cm. Three days after birth convulsions developed, at that time the CSF (lumbar) was xanthochromic (bilirubin, 0.1 mg per 100 ml) and Pándy's test was positive. From the second week on the infant's head began to grow fast. In the third week the head circumference was 34 cm. During that period the infant suffered from dyspepsia, pharyngitis and conjunctivitis. The stools were repeatedly negative bacteriologically. There was no fever. After the third week the CSF was water-clear, with a cell count of $^4/_3$ and Pándy's test ++. In the seventh week the head circumference was 41 cm, and continued to increase at a dramatic rate. At the age of 3 months it was 50 cm. Ventricular punctures were carried out at regular intervals, withdrawing 100 to 150, then 500 ml of CSF because lumbar punctures failed to yield sufficient volumes of CSF to diminish intracranial pressure. At the age of 7 months the circumference of the skull was 67 cm, lumbar CSF pressure was 150 mm water, ventricular pressure over 300 mm water. Methylene blue injected into the ventricle did not appear in the lumbar CSF. In the lumbar CSF Pándy's test was +, in the ventricular CSF, negative. Occlusive hydrocephalus was diagnosed. The occipital part of the skull was particularly well developed, the external occipital protuberances took place higher than usual. In view of the characteristic overdevelopment of the posterior scala, the occlusive hydrocephalus was considered a Dandy—Walker syndrome, based on a developmental abnormality. The poor condition and chronic bronchitis of the patient contraindicated neurosurgery.

The infant's condition continued to deteriorate, the baby weighed 5550 g, the circumference of the skull reached 70.5 cm. Then pressure by the enormous head led to the development of a CSF fistula. After the escape of CSF the central part of the skull showed a deep impression 6×9 cm in size. Two days later the infant died, at the age of 9 months.

b) *Gross and Microscopic Anatomy*

Prior to autopsy, the ventricular system was filled up repeatedly with 10 per cent formalin through a cannula introduced into the lateral ventricles, then after fixing for 24 hours the formalin was replaced by paraffin. The paraffin cast filling out the ventricles and together with it the brain tissue around it were cut up into 15 mm thick sections (Fig. 1). It was found that neither the formalin, nor the paraffin had entered the fourth ventricle, the paraffin filled out only the two enormously dilated lateral ventricles and the third ventricle, and entered a fist-sized subarachnoidal cyst through an opening in the posterior part of the third ventricle; the cyst was located superior to the vermis and the cerebellar hemispheres (Fig. 3 and Fig. 26). The third ventricle was dilated, the optic recess, the infundibulum and the infundibular recess could not be recognized separately, they were confluent, the floor of the third ventricle ballooned out between the peduncles, the mammillary bodies could not be detected. In the posterior part of the third ventricle, where the aqueduct of Sylvius ought to have begun, there was a bulge ending blindly, over it there was brain tissue about $^1/_2$ cm thick, corresponding to the posterior commissure, and above that an opening, 12×12 mm in size, in the area corresponding to the pineal recess in the posterior wall of the third ventricle, that communicated with the fist-sized subarachnoidal cyst. The compressed pineal body was near to that opening, above the cyst. The cyst flattened the corpora quadrigemina, whose shape could not be recognized. In the transversal sections it is visible that not far below the area corresponding to that of the rostral collicle the cavity of the fourth ventricle began blindly (Fig. 2). The fourth ventricle was not dilated, the

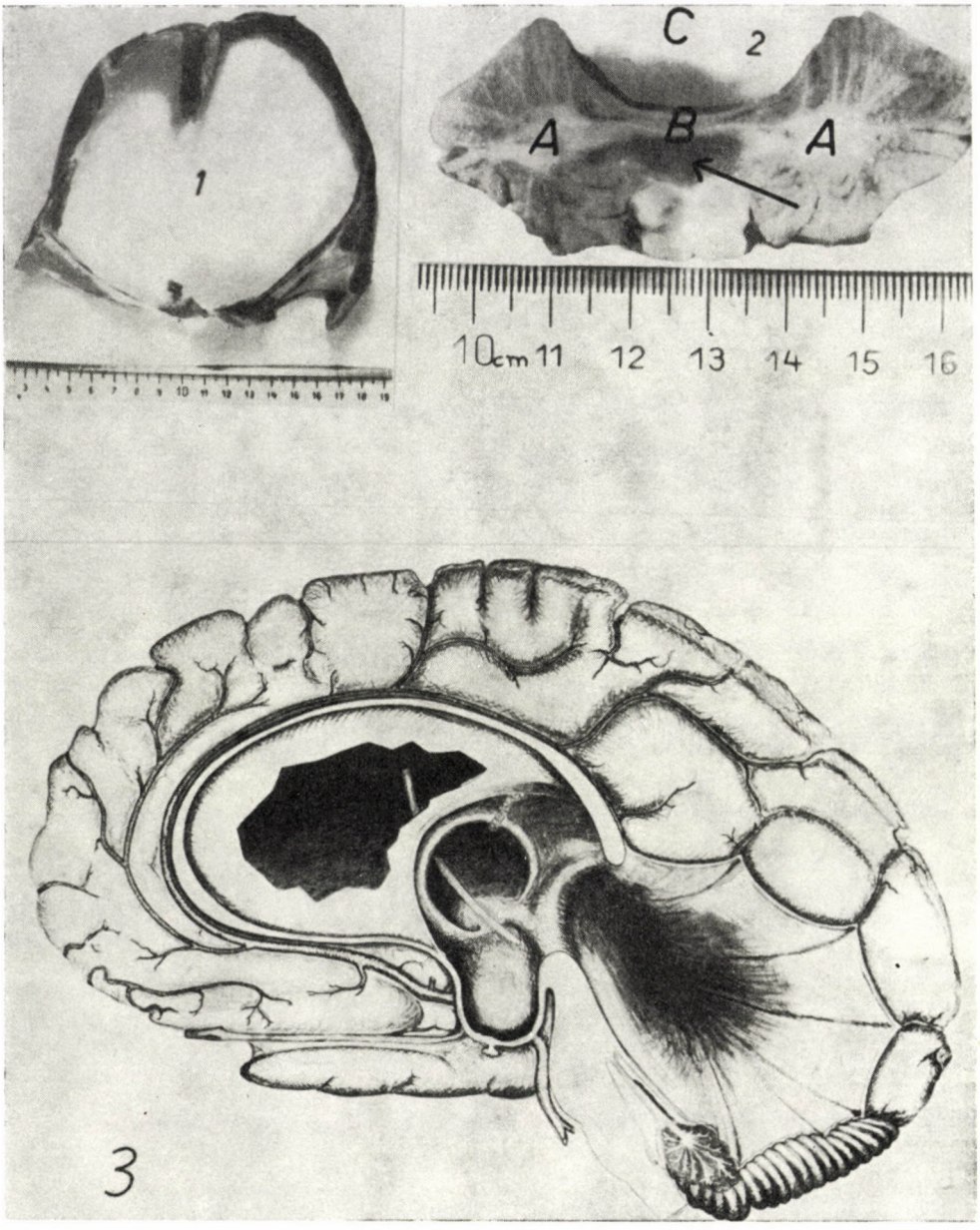


FIG. 1. Case 1. Frontal section of the brain, at the middle of the thalamus. FIG. 2. Case 1. Transversal section at the level of the centre of the oliva; caudal appearance. A: cerebellar hemispheres, B: flattened vermis, C: site of supracerebellar subarachnoidal cyst. The arrow points at the area in which the fourth ventricle begins blindly. FIG. 3. Case 1. Diagram of the sagittal section of the brain, reconstructed on the basis of the frontal slices. A probe is inserted into the dilated Magendie's foramen. Note the dilatation of the lower part of the third ventricle and the location of the supracerebellar subarachnoidal cyst

two lateral ventricles and the third ventricle had a combined capacity of 4530 ml, as determined from the paraffin cast. The corpus callosum was transparent, membrane-like, the cerebral matter was 1 to 2 mm thick. In the posterior arch of the atlas there was an about 6 mm wide dehiscence in the midline, the cerebellar tonsils were herniated, Magendie's foramen was about 3 mm in diameter, the vermis was flat. The diagram of the reconstructed sagittal section of the brain is shown in Fig. 3 and Fig. 4.

In the other organs no pathological changes were found except for a marked anaemia.

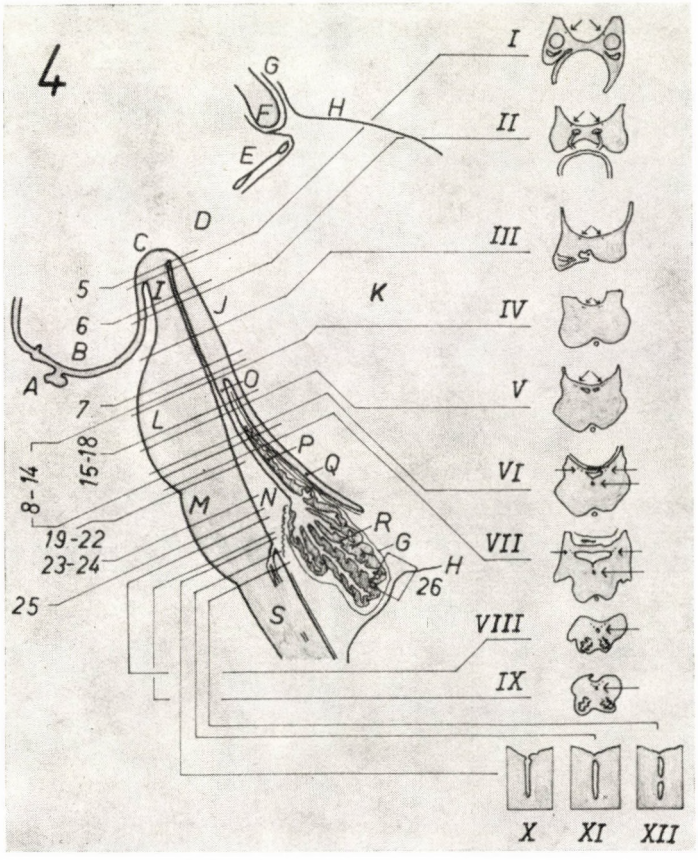
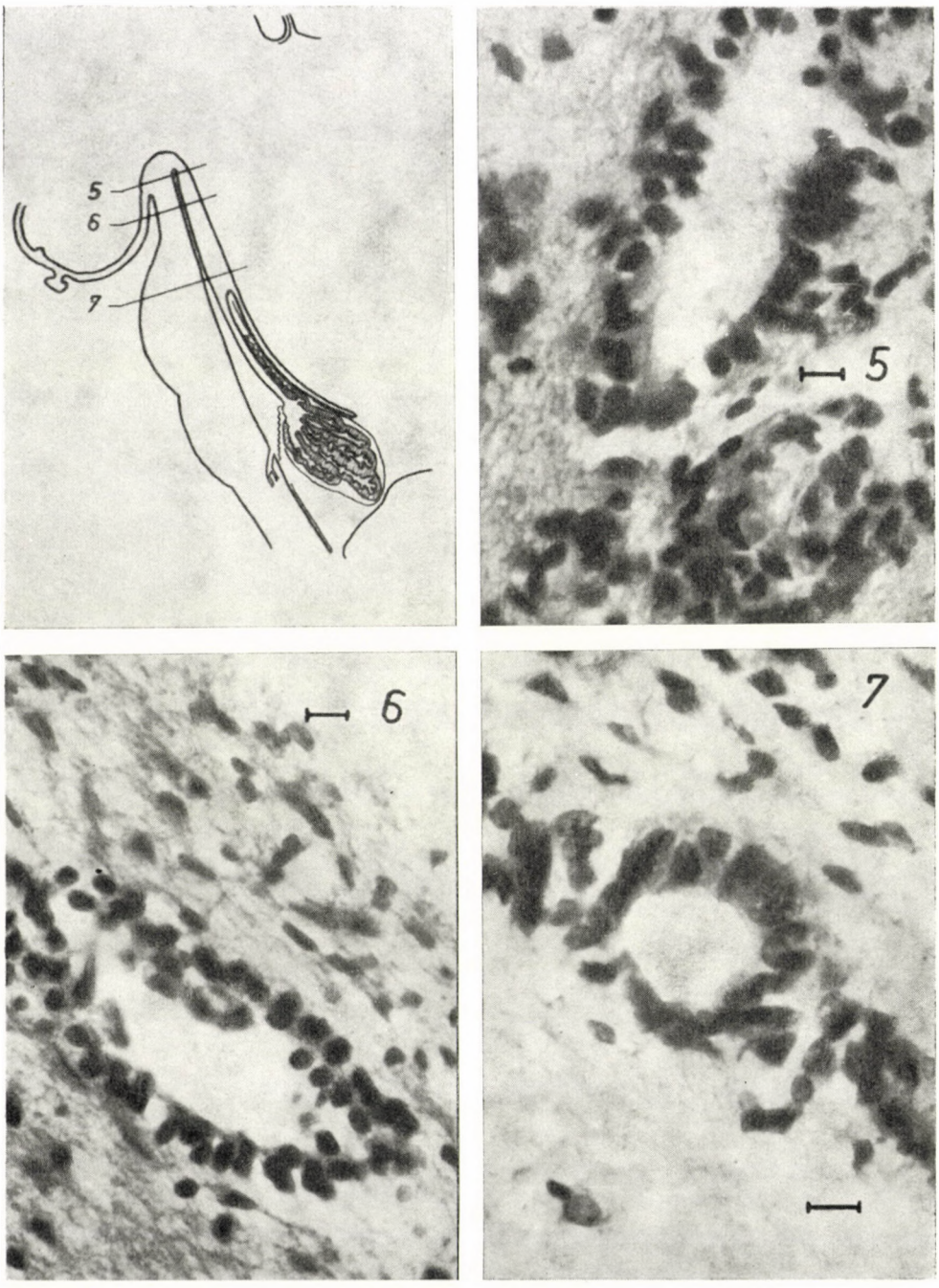


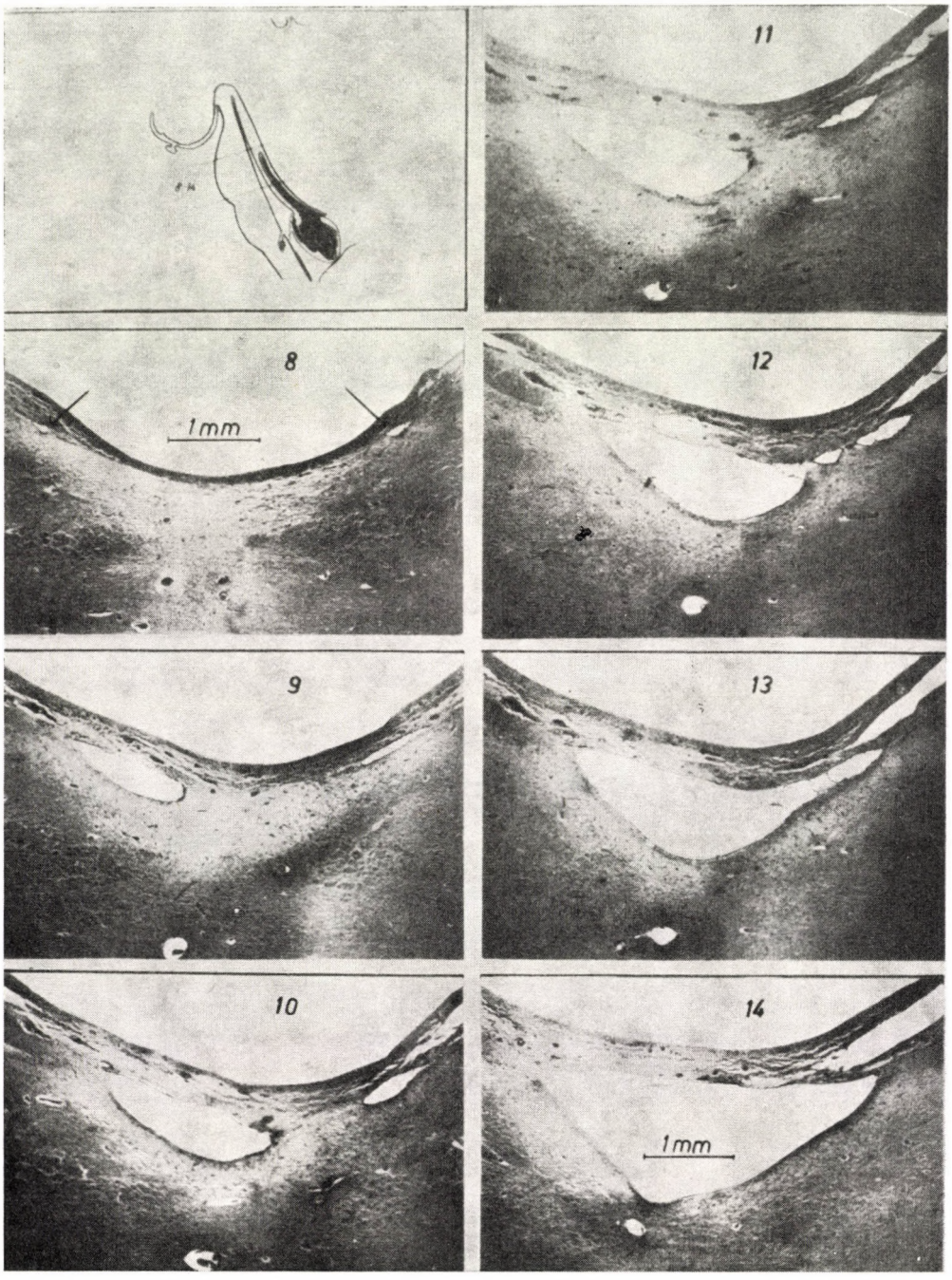
FIG. 4. Case 1. Diagrammatic representation of the brain stem, in sagittal section. A: Pituitary. — B: Infundibulum, dilated. — C: Area corresponding to the posterior commissure. — D: Opening leading from the third ventricle into the supracerebellar subarachnoidal cyst. — E: Pineal body. — F: Splenium corporis callosi. — G: Pia mater encephali. — H: Arachnoid. — I: Cerebral peduncle. — J: Lamina tecti. — K: Subarachnoidal cyst. — L: Pons. — M: Oblong medulla. — N: Fourth ventricle. — O: Anterior velum medullare. — P: Cerebellar lingula. — Q: Residue of the ruptured posterior wall of the third ventricle. — R: Vermis. — S: Spinal medulla

Arabic numerals are those of the microphotograms shown in Figs. 5 to 26. Figures I to IX show the cross sections of the single segments of the brain stem, in 1 to $^2/_3$ the original size. The arrows point at the aqueducts. Figures X to XII show the beginning of the central canal and its bifurcation, as seen under the magnifying glass

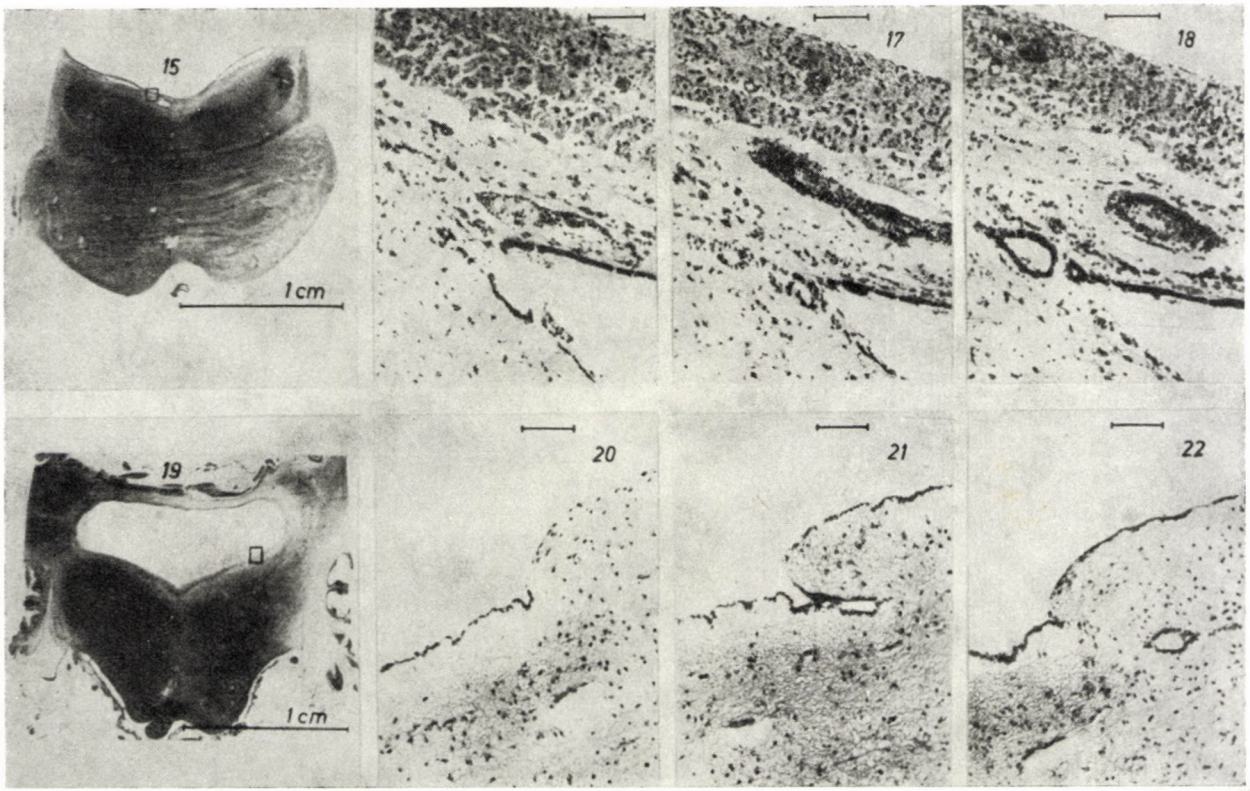


FIGS. 5, 6 and 7. Case 1. Aqueductules from various segments. In Fig. 5, below: group of ependymal cells without lumen

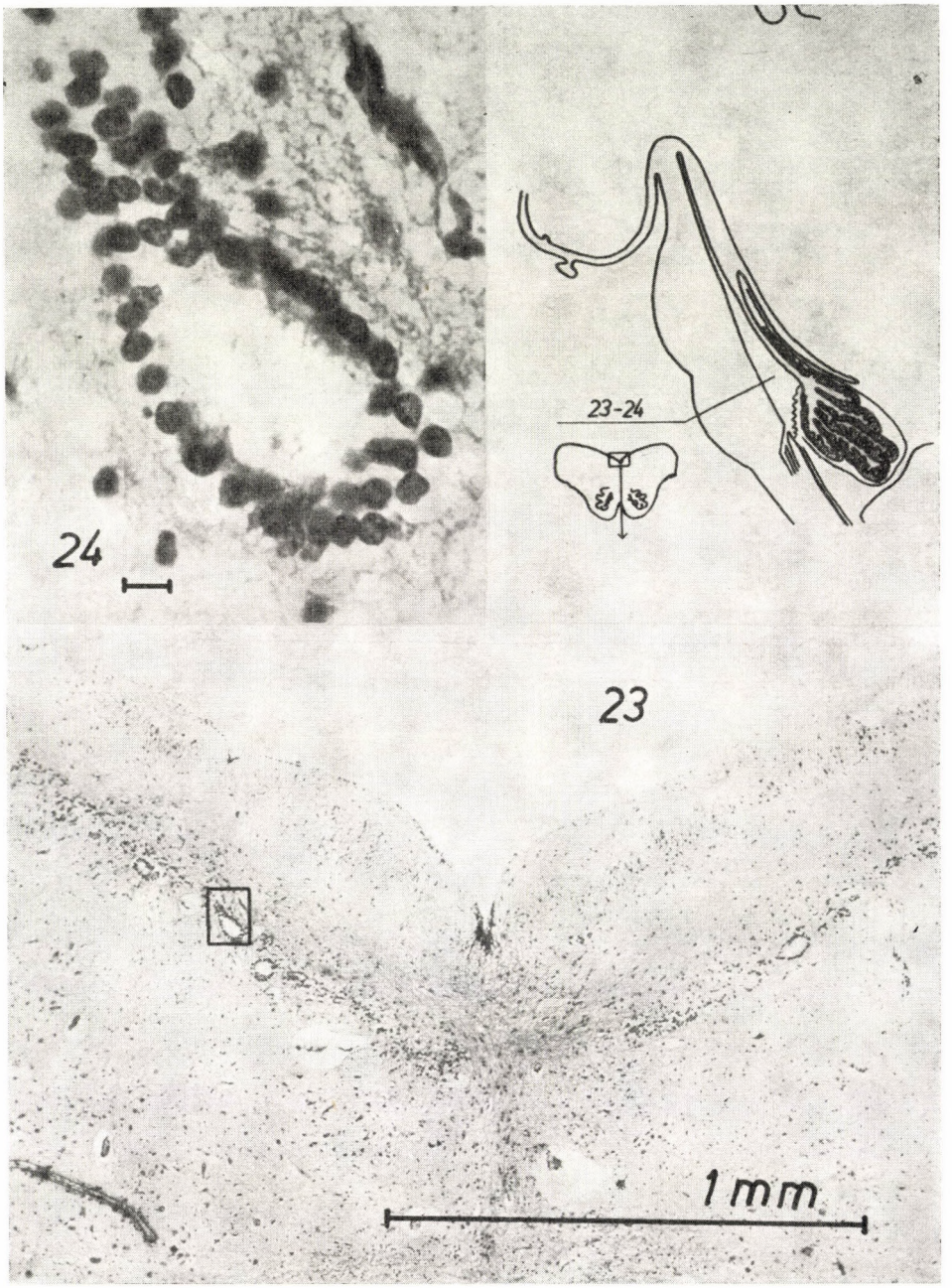
Staining: haematoxylin-eosin. Lens: ×40 achr. Eyepiece: Homal VI. Extension with Figs. 5 and 6, 55 mm; with Fig. 7, 75 mm. Subsequent paper magnification, ×3. Scale, 10 microns



FIGS. 8 to 14. Case 1. Transition of aqueducts into the fourth ventricle. The arrows in Fig. 8 point to aqueducts. Haematoxylin-eosin. Magnification ×13. Scale, 1 mm



FIGS. 15 to 18. Case 1. Origin of aqueducts from the corners of the fourth ventricle, from ependyma-deficient areas. FIGS. 19 to 22. Case 1. Origin of aqueductules from the floor of the fourth ventricle (upper part of Fig. 20). Haematoxylin-eosin. Magnification: Figs. 15 and 19, ×2.6; Figs. 16—18 and 20—22: Lens, ×10 apochr, Eyepiece, Homal VI. Extension, 55 mm, subsequent paper magnification, ×2. Scale, 100 microns



FIGS. 23 and 24. Case 1. Multitude of aqueducts in the floor of the fourth ventricle. Haematoxylin-eosin. Magnification: Fig. 23, Mikrotar ×63. Fig. 24, Lens, achr. ×40. Eyepiece, Homal VI. Extension 75 mm. Subsequent paper magnification, ×3. Scale, 10 microns

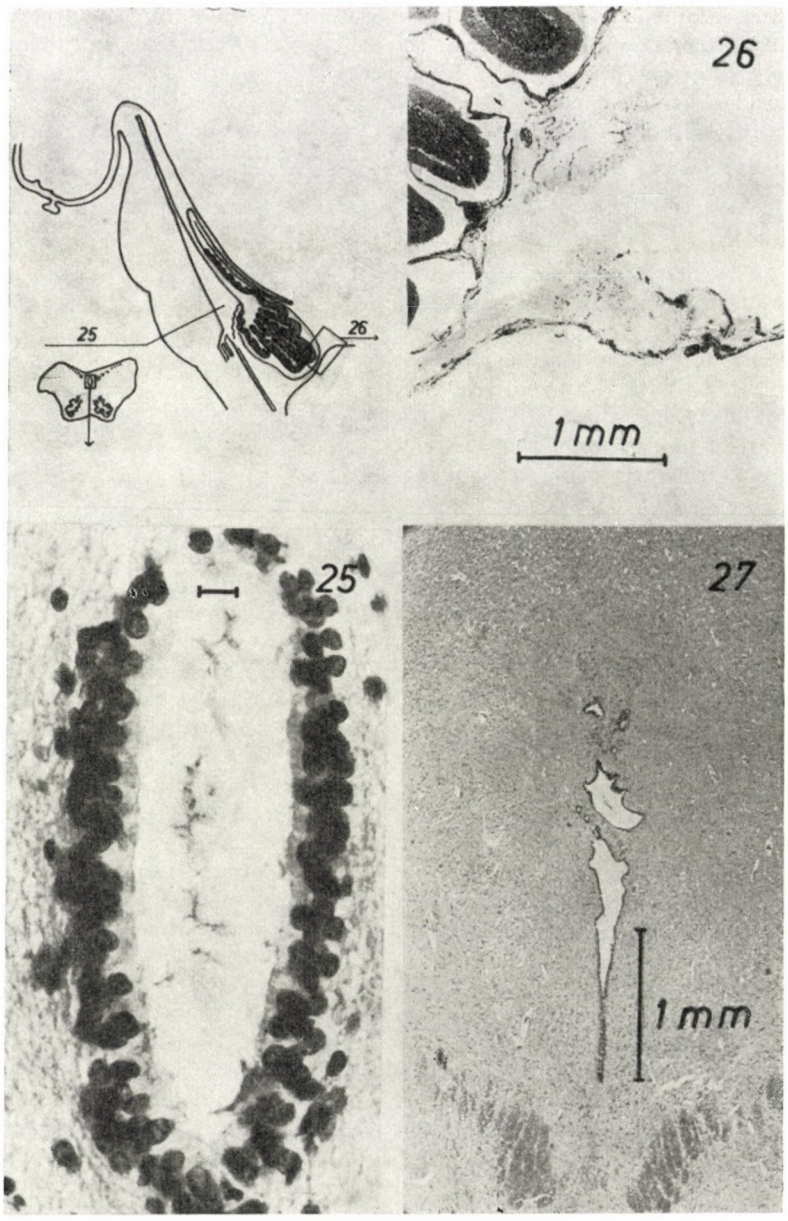


FIG. 25. Case 1. Canalis centralis-like canal from the floor of the fourth ventricle, from the area of the centre of the oliva. Haematoxylin-eosin. Lens, achr. ×40; eyepiece, Homal VI. Extension 75 mm, subsequent paper magnification, ×3. Scale, 10 microns.

FIG. 26. Case 1. Margin of supracerebellar subarachnoidal cyst. Haematoxylin-eosin. Magnification: Mikrotar ×24.

FIG. 27. Case 2. Aqueduct at its narrowest point. Haematoxylin-eosin. Magnification: Mikrotar ×25

The mesencephalon, the metencephalon (without the cerebellar hemispheres), the myelencephalon and the uppermost part of the cervical cord were cut up into serial sections; furthermore, 1 section was made from each cervical segment. The sections were stained with haematoxylin-eosin, van Gieson's, Nissl's, Holzer's and Bielschowsky's stains. In the course of the histological studies in some transversal sections numerous microscopical as well as lumen-free groups of ependymal cells and bundles were found. The aqueducts did not communicate with the third ventricle, they began blindly 1.8 mm caudally from the third ventricular surface of the posterior commissure, in the form of small-calibre canalicules (Figs. 5 to 7), ramifying several times running downwards. Some of them ended blindly, others communicated with each other, still others opened into the fourth ventricle (Figs. 8 to 14). Numerous ductules ran in the floor of the fourth ventricle, in its entire length and width (Figs. 23 to 25). Some of these originated from the posterior commissure (see above), while others from the fourth ventricle (Figs. 15 to 22). Every canalicule opened into the fourth ventricle, none of them continued in the central canal of the cord. The central canal started in the proper place, a short distance away it showed a bifurcation, then at C VII it reunited again. In the floor of the fourth ventricle ependymal defects with bulging glial tissue were found in many areas. At the margin of the defect the ependyma was penetrating into the tissue and continued either in lumenless bundles or lead to the ductules (Figs. 19 to 22). Around the ductules glial tissue and occasional nerve cells could be found.

There was no evidence of tumour or inflammation, the pattern was that of an atresia and forking based on a developmental abnormality, in which instead of the suitably wide aqueduct capable of permitting the flow of CSF numerous aqueductules of very narrow calibre, showing forking, ending blindly eventually, and beginning blindly, could be found, together with lumenless ependymal bundles.

Case 2.

a) *Clinical Data*

R. M., a female infant was born from the first pregnancy of her mother. Weight at birth, 1520 g; circumference of the skull 33.5 cm; wide fonticles. After birth the circumference of the head increased in size at a fast rate. The infant failed to develop and at the age of 3 months died of pneumonia. At that time she weighed 2300 g, the circumference of the head was 45 cm.

b) *Gross and Microscopic Anatomy*

Prior to autopsy the lateral ventricles were punctured, the CSF was replaced by 10 per cent formalin. The brain was thus fixed *in loco* for 24 hours, then it was removed, fixed in 10 per cent formalin for 8 days, then cut up into frontal slices under water. The brain substance measured merely 2 to 4 mm in some areas. The lateral ventricles, as well as the third ventricle were extremely dilated. In the posterior wall of to the third ventricle, over the posterior commissure, there was a round opening 20 mm in diameter, leading into a fist-sized subarachnoidal cyst on the dorsal aspect of the cerebellar vermis and the cerebellar hemispheres. The origin of the aqueduct was not visible, the fourth ventricle was not dilated, it began blindly at the level of the rostral collicles. In the areas below the cyst, the collicles, the vermis and the cerebellar hemispheres were somewhat flat.

In the other organs there was bilateral bronchopneumonia and degeneration of the heart, liver and kidneys.

In the serial sections cut from the mesencephalon (staining: haematoxylin-eosin, van Gieson, Nissl, Holzer, Bielschowsky) we found the site at which the narrow and forking aqueduct started from the third ventricle. There was no atresia, only stenosis and forking (Fig. 27).

Discussion

On the basis of the excessive occlusive hydrocephalus, the exaggerated development of the occiput, the high position of the external occipital protuberance, we tentatively diagnosed *in vivo* Dandy—Walker's syndrome, a cyst-like dilatation of the fourth ventricle (congenital occlusion of Magendie's foramen). At autopsy, however, instead of a cyst-like dilatation of the fourth ventricle, a subarachnoidal cyst on the dorsal surface of the cerebellar vermis and the cerebellar hemispheres was found. The posterior wall of the third ventricle showed a diverticulum-like formation. This had ruptured under the pressure of the CSF, creating a spontaneous ventriculostomy. The CSF escaped into the infratentorial subarachnoidal space had given rise to the cyst which caused excessive growth of the skull, and especially of the occiput; this, *in vivo*, imitated the skull deformity described by Dandy and Walker.

The gross changes were identical in the two cases, both the atresia and the stenosis gave rise to the same type of hydrocephalus. In view of this we feel justified to assume that in Case 2 the stenosis failed just as well in draining away the CSF, as did the atresia in Case 1, i.e. from the functional point of view the stenosis of the aqueduct in Case 2. should be considered an atresia.

Owing to the variability of the size of the aqueduct, it is difficult to judge on the basis of its calibre alone whether or not there is stenosis. According to Turkewitsch [19] in a foetus 35 cm long S. S. L. the aqueduct is narrowest near the genu. Here the lumen is triangular in shape, with the base 0.5 mm, the height 2.1. mm; the two sides are equal in length and slightly arched. According to our calculations the surface area measures about 0.54 sq. mm. Studies of the aqueduct have been carried out in infants and adults without hydrocephalus by Spiller [18] in 38 cases, by Beckett et al. [2] in 50, by Woollam and Millen [20] in 14, as well as by Flyger and Hjelmquist [6] in 22 cases. Every author found extreme variations both in shape and calibre. The surface area of the narrowest aqueduct not accompanied by hydrocephalus observed by Beckett et al. [2] was 0.9 sq. mm. in the transversal section (corrected according to Woollam and Millen [20]). The narrowest aqueduct measured by Woollam and Millen [20] measured 0.20 sq.mm, that found by Flyger and Hjelmquist [6] in an adult without hydrocephalus measured 0.40 sq. mm, the widest 9.84 sq. mm, and in two newborns values of 0.54 and 0.84 sq. mm were found. Bickers and Adams [3] described a case of stenosis associated with hydrocephalus. We measured the lumen of the aqueduct in their figures and found it between 0.16 and 0.20 sq. mm. In our two cases of stenosis there were two lumina at the narrowest point of the aqueduct; the sum of their surface areas was 0.14 sq. mm (Fig. 27). The narrowest lumen (0.40 sq. mm)

observed by FLYGER and HJELMQUIST [6] was merely $^1/_{25}$ of the widest one they found and yet there was no hydrocephalus in the former case. The lumina measuring 0.14, 0.16 and 0.20 sq. mm, amounting to $^1/_3$ to $^1/_2$ of the 0.40 sq. mm aqueduct were accompanied by hydrocephalus (our own case 2, and the case of BICKERS and ADAMS [3]), whereas no hydrocephalus was found by WOOLLAM and MILLEN [20] with an aqueduct measuring 0.20 sq. mm.

From the above it follows that the diagnosis of aqueductal stenosis can be made only when the lumen of the aqueduct is narrower than 0.20 sq. mm and at the same time a hydrocephalus of the appropriate type is present, i.e. when the consequences of stenosis are also observable. A division of the aqueduct alongside its stenosis or atresia has been mentioned by most of the authors. Suggesting this to be a characteristic accompanying change, RUSSEL proposed the term of forking instead of stenosis or atresia. The branching of the aqueduct occurs not exclusively with stenosis or atresia; even under normal conditions accessory aqueductules and lumenless groups and bundles of ependymal cells may be present beside the main aqueduct [2, 7], that originate mostly from such areas of the aqueduct in which the ependyma is absent and the glia bulges into the lumen. This phenomenon could be observed in our Case 1 (Figs. 15 to 22). Thus, forking is not necessarily a pathological phenomenon.

It has been suggested that the developmental disorder leading to aqueductal stenosis or atresia is due to some intrauterine infection retarding the growth of the mesencephalic anlage, or to an involution of the aqueduct analogous to the obliteration of the central canal of the cord [18]. In human embryos, foetuses and premature infants FLYGER and HJELMQUIST [7] found under the rostral part of the superior collicles, dorsal to the aqueduct, circumscribed groups of ependymal cells and cells whose nature could not be determined precisely, to which they have ascribed an important role in the aetiology of congenital atresia or stenosis.

In this relation the problem of heredity and repeated occurrence emerges. BICKERS and ADAMS [3] observed a newborn with congenital hydrocephalus and stenosis of the aqueduct. The infant died shortly after birth, just like his two brothers, born also with hydrocephalus. The mother was a healthy woman of 30, among whose 9 siblings there were 3 healthy females, 1 healthy male, one still-born normal male and 4 still-born males with congenital hydrocephalus. It would therefore appear that the factor at issue is a male sex-linked, female-transferred hereditary defect. KOVÁTS [11] reported on 3 hydrocephalic sisters with club foot, born from the marriage of first cousins; from the fourth pregnancy a mature, normal girl was born. This speaks against the male sex-linkedness, and suggests an aetiological role of consanguinity. In our own

cases the parents were healthy; hydrocephalus, hereditary diseases had not occurred in the family, there was no evidence of consanguinity. Both our patients were females, the parents of Case 1 gave birth since then to a normal male, who now is one year old. In the light of these facts we do not think it proved that heredity, sex-linkedness or consanguinity have any major role in the aetiology of hydrocephalus caused by aqueductal stenosis or atresia.

Conclusions

(i) The skull deformity described by Dandy and Walker (congenital occlusion of Magendie's foramen) can be caused not only by a ballooning of the fourth ventricle, but also by other changes in the posterior scala, influencing the growth of the bones of the skull, thus for example a supracerebellar cyst caused by aqueductal stenosis or atresia and the spontaneous ventriculostomy of the posterior wall of the third ventricle.

(ii) We have observed a case of aqueductal atresia that showed forking at a more caudal level. Thus, atresia of the aqueduct and forking may occur together.

(iii) In our opinion the histological diagnosis of stenosis of the aqueduct can be made only when the lumen of the aqueduct is narrower than 0.20 sq. mm, and simultaneously there is also hydrocephalus extending to the lateral ventricles and the third ventricle.

(iv) The parents of children with atresia or stenosis of the aqueduct and hydrocephalus may later give birth to normal children. Thus, it cannot be proved that atresia or stenosis of the aqueduct would be due to hereditary, sex-linked or consanguinity-induced factors.

Summary

Two cases of congenital hydrocephalus were observed with stenosis and forking of the aqueduct. In one case there was also aqueductal atresia. In both cases a supracerebellar, subarachnoidal cyst arising as a result of the spontaneous rupture of the posterior wall of the third ventricle could be detected. The latter change was responsible for the skull defority, observable also in Dandy—Walker's syndrome.

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