SMALL VASCULAR MALFORMATIONS OF THE BRAIN:
THEIR RELATIONSHIP TO UNEXPECTED DEATH,
HYDROCEPHALUS AND MENTAL DEFICIENCY*†

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Small vascular malformations of the brain have been increasingly often recognized in recent years as possible sources of intracranial hemorrhage, especially in young people (1–6). There are many other problems in connection with these lesions. It is the purpose of this paper to illustrate by means of case studies some of the local results of these malformations and also to discuss the multiple form and its relationship to generalized vascular disturbance of the brain. The association of these malformations with dysgeneses of the nervous tissue will also be discussed.

SUBARACHNOID HEMORRHAGE

Subarachnoid hemorrhage due to rupture of a small vascular malformation is found with some frequency in children and young adults who have died unexpectedly. It is usually impossible to learn of any premonitory signs or symptoms, even though relatives and friends are carefully questioned.

In these cases, the neuropathologist is faced with the difficult problem of finding a minute lesion in a large amount of blood. This blood may appear on inspection to be completely confined to the subarachnoid space when the malformation is adjacent to the pia. If so, careful inspection of thin slices of the brain with a hand lens may be the simplest means of identifying the region of the malformation; final proof must necessarily be by microscopic section. Quite often, however, the malformation is located somewhat deeper. In this event, an intracerebral hematoma is produced. As this enlarges, it reaches and ruptures into the subarachnoid space. Margolis et al. (2) have described 6 such instances. It is noteworthy that their case histories indicate an interval of a few days between onset and death. During this time, the bleeding may have been wholly intracerebral, the terminal event being rupture into the subarachnoid space. If there has been a large amount of bleeding into the tissues of the brain, it is well to remove the blood gently, bit by bit, so as to expose the margin of the lesion (1). It is in this region that the vascular malformation is most likely to be found.

The malformations discussed here are minute, seldom being over 5 mm. in diameter. In the gross, they appear gray, or yellow if there has been a little bleed-

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ing some time previously. Occasionally, abnormal and distended vessels are identified grossly within the area; more often, their presence is established only by histologic study.

Under the microscope, there is a group of rather large vessels which are irregular in shape. Most walls are thin, but from place to place fibromuscular cushions project from the wall. These make a localized thickening in contrast to the delicacy of the rest of the vessel. The vessels may appear as arteries or veins, or (more often) they are too abnormal for classification as either with certainty. In the present material, the cerebral tissue between the vessels showed no definite disgenesis, the only abnormality being a slight increase in astrocytes.

In this group of vascular malformations, the absence of warning signs and symptoms usually precludes preventive therapy, in spite of their small size and their accessible locations for surgical attack. While angiography can demonstrate many other forms of vascular malformation, it is doubtful that this procedure can identify those as small as the ones here discussed.

Fig. 1. Case 1. Vascular malformation in the cerebrum. Variations in structure in the walls of the vessels are shown. About half of the entire lesion is included in the field. Gallo-cyanin-van Gieson, × 90.
CASE REPORT

Case 1. (From Dr. Richard Ford, Boston). A white male, 17 years of age, died suddenly on the street. So far as his relatives knew, he had been in excellent health.

Post Mortem Findings: At autopsy, massive subarachnoid hemorrhage was found. After the blood had been removed from the surface, the convolutions appeared normal, except in an area 0.5 cm in diameter on the lateral aspect of the left frontal lobe. This region was yellowish gray and a small amount of blood was present within the lesion.

In microscopic sections, the area was composed chiefly of large blood vessels. These were irregular in outline and had thin walls, except for a few segments where fibromuscular cushions jutted out from the intima (fig. 1). The tissue intervening between the blood vessels was disturbed from the usual pattern of gray matter only by a moderate increase in astrocytes. A few hemosiderin-filled phagocytes were present within it. Other portions of the brain were free from developmental abnormalities.

DISCUSSION

This patient illustrates the difficulty in instituting diagnostic or operative procedures in instances of small vascular malformations. Occasionally, the lesion, even though minute, occurs in a region where there are diagnostic signs. Surgical extirpation may then be accomplished with considerable assurance of permanent cure.

Fig. 2. Case 2. Vascular malformation with Jacksonian epilepsy. Fibrous and hyaline areas are continuous with thin segments in the vessel walls. Gallocyanin-van Gieson, X 130.
Case 2. (From Dr. Eben Alexander). An 8 year old Indian boy had 2 focal seizures beginning in the right arm approximately 2 months before admission to the hospital. Left frontal headache occurred just before the onset of seizures but had not persisted. The boy had not done quite as well as before in school during the previous year. No abnormal neurological findings were discovered, but a pneumoencephalogram showed a right ventricular shift of 8 mm., with some compression of the left frontal horn. A left arteriogram demonstrated slight upward displacement of the anterior cerebral artery but no abnormal vessels were seen.

At operation, there was a discolored area 4 mm. in diameter in the second left frontal convolution 4 cm. anterior to the Rolandic fissure. That area was bluish and gelatinous. No further lesions were found by exploration with a ventricular needle.

Two years after operation, the patient was well and had had no seizures. He had, however, received anticonvulsant medication. An electroencephalogram was entirely normal.

Microscopic Examination showed a group of anomalous vessels extending through relatively well preserved cerebral tissue (fig. 2). Some vessels had thick hyaline walls, often with plaques of fibrous tissue beneath the intimal surface. In continuity with such vessels, there were others with very thin walls.

INTRAVENTRICULAR HEMORRHAGE

Small vascular malformations may occur near the ependymal surface. When they rupture, the ventricular system quickly fills with blood. Since the bleeding is initiated within the cerebral substance, the intraventricular hemorrhage may be combined with hemorrhage into the subarachnoid space.

In most instances in which the bleeding is wholly or predominantly intraventricular, the malformations are less focal than those nearer the surface of the brain. There is an abnormal vascular pattern in a comparatively small area, often with disordered organization of the nervous tissue restricted to the region of malformed vessels. It is suggested that the internal cerebral vein and its tributaries are especially often involved in lesions of this type.

Fig. 3. Case 3. Crinkled and deeply infolded vein in a region of abnormal vessels adjacent to the third ventricle. Galloecyanin-van Gieson, X 160.
CASE REPORT

Case 3. (From Dr. Lall G. Montgomery). A 13 year old boy complained of headaches at intervals of 2 months over a period of about 2 years. Otherwise, he was healthy, active, and very good in school. On the day of his death, severe pain developed in the right side of his head. He was conscious for half an hour, then became comatose. There were tremulous twitchings but no definite seizures developed. Death occurred 4½ hours after the onset of the severe headache.

Post Mortem Findings: At autopsy, the only external change was a small amount of blood in the subarachnoid space at the base of the brain. When the brain was cut after fixation, the entire ventricular system was found to be full of blood. On examination of the walls of the ventricles an area of hemorrhage was found adjacent to the third ventricle on its right. This extended into the corpus striatum and septum pellucidum with rupture through the ependyma and in the region of the medial orbital convolutions. On microscopic study, abnormal veins of large caliber were found in the region of hemorrhage and adjacent parts of the corpus striatum. Some of these had deeply infolded and crinkled walls (fig. 3). In the wall of one such vein and around it, there were polymorphonuclear leukocytes in considerable numbers, suggesting that this was the point of rupture. There were groups of small dysgenetic foci of nervous tissue, especially near the septum pellucidum. In the subarachnoid space of the medial orbital convolutions, there were further vascular malformations, especially of the arteries. These vessels showed thick walls with fibromuscular cushions (fig. 4) continuous with thin segments. In all other regions, the brain was free from vascular malformations and nervous tissue dysgenesis.
HYDROCEPHALUS

In the patients so far described, small vascular malformations produced symptoms by rupture (cases 1 and 3) or by involvement of an area associated with motor activity (case 2). A rather different mechanism is found when a small malformation occurs beneath the ependyma of the aqueduct of Sylvius. If the abnormal vessels are aggregated into a relatively discrete focus in this location, they may wholly or partially obstruct the aqueduct. Hydrocephalus then results from mechanical effects on a relatively small but vital anatomical structure.

Routine study of many sections of the neuraxis in unselected brains indicates that some variations from the usual patterns of vascular structure and distribution are frequent in the subependymal region near the third ventricle. Similar variations are often found also beneath the floor of the fourth ventricle, where they may be aberrations of the normal process by which growing vasculature undermines the neuroepithelium of the myelencephalic rhombic lip.

In the vascular malformations obstructing the aqueduct, the blood channels are predominantly venous in structure. Some of them bulge into the aqueduct beneath an attenuated ependymal layer. It is by this process that obstruction takes place. Between the vessels, there is usually nervous tissue with minimal gliosis and with neurons in normal or somewhat abnormal pattern. Nests of ependymal cells, not always fully mature, are frequent. These findings would suggest a focal malformation of many tissues in a small region rather than abnormal development of blood vessels only.

When hydrocephalus results from a single area of malformation predominantly vascular, it may reach an extreme degree. At the same time, the enlargement of the head may be delayed, or not clearly apparent, for a month or longer. Clinical identification of the cause of the hydrocephalus in these patients is difficult or impossible with the diagnostic methods now available.

CASE REPORT

Case 4. (From Muscatatuck State School, Mr. A. E. Sasser Superintendent, Dr. Hans Meyer, Clinical Director). A male negro baby appeared normal until the age of 1 month; after this time the head enlarged rapidly. Its circumference reached 72 ems. at the age of 7 months, when death resulted from bronchopneumonia.

Post Mortem Findings: At autopsy, there was advanced hydrocephalus, the cerebral mantle averaging 1 cm. in thickness. The aqueduct was obstructed by a group of very thin-walled vascular channels (fig. 5). These represented a concentration of abnormal vessels which were also present in the region of the area postrema and in the tectum of the midbrain. Well preserved neurons were found between the vessels, and there was no detected astrocytosis in the region. Vessels elsewhere in the brain were normal. Other changes in the central nervous system were secondary to the mechanical effects of the hydrocephalus. No other foci of vascular malformation were found in other parts of the nervous system or in the extracranial viscera.

MENTAL DEFICIENCY

In certain patients with a history of mental deficiency, small focal vascular malformations are found scattered through many parts of the brain. On further
study, it is apparent that these foci are grossly visible parts of an extensive anomalous development of cerebral vasculature. While mental deficiency has usually been found in such patients in the present series and in those reported by others, it would seem very doubtful that the vascular anomalies *per se* are to be correlated with this status. The abnormal vascular structures could be interpreted as one part of a disturbance in growth of cerebral tissue.

These small, multiple, vascular malformations may be found in any part of the brain but are relatively more frequent in the white matter of the cerebral convolutions, in the tegmentum of the caudal mesencephalic and rostral pontine levels, in the basis pontis, in the central cerebellar white matter and among the cerebellar folia.

When they occur in the cerebral white matter, there is often considerable disorganization of the overlying cortex. This disorganization may be apparent only as slight variation in neuronal arrangement or, in more extensive instances, by the presence of a deep sulcus extending to the site of the vascular anomaly.

On the other hand, one of these multiple malformations may impinge upon the aqueduct and cause hydrocephalus by the same mechanism as an isolated malformation in this region (case 4). If the vascular abnormality is placed elsewhere in the neuraxis, there is usually little or no effect on nuclear organization. Extensive examples may extend across the mesencephalic tegmentum without apparent disturbance of neuronal distribution.

Vascular anomalies in the cerebellar central white matter ordinarily produce no appreciable disturbance in cerebellar growth or localizing clinical symptoms. Anomalous vessels may involve a group of cerebellar folia and produce considerable disorganization of the cortical pattern. This results in a mixture of areas of molecular layer, granule neurons and isolated Purkinje neurons about
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FIG. 7. Case 4. Vascular malformation protruding into the aqueduct of Sylvius. There are abnormal vessels along the wall of the aqueduct in addition to the larger mass. Gallocyanin-van Gieson, X 80.

the anomalous vessels. The size and general pattern of the involved folia is little disturbed.

These small, multiple vascular malformations are easily seen in the course of gross dissection of the brain. When they are so demonstrated in an otherwise well developed brain of a patient with mental deficiency, it is tempting to relate the malformations and the clinical disturbance directly. This procedure is open to very serious objection. In the present material, it would seem that the vascular abnormalities may exert effects mechanically (hydrocephalus), or that they may be the easily discernible part of a more generalized developmental disturbance. Even though malformation of the nervous tissue is not demonstrated by present methods, any direct correlation of mental deficiency and the small vascular malformations is at best facile.

CASE REPORT

Case 5. (From Central State Hospital, Dr. C. L. Williams Superintendent). A white woman, who died at the age of 48 years, had been in a mental institution since the age of 19.
years because of mental deficiency. Mental progress had been slow from early childhood and she had reached the fourth grade in school at the age of 17 years. She then left school for the birth of an illegitimate child; at the time of the birth of another 2 years later, mental deficiency was found so severe that institutional care was essential. A careful restudy was done at the age of 39 years. The patient was mentally very retarded and had very poor memory. Blindness had come on suddenly, and there was bilateral optic atrophy, nystagmus and reactionless pupils. Because of a deep excavation in the region of the sella turcica, demonstrated by X-ray, a clinical diagnosis of pituitary adenoma was made. She continued in much the same state for 9 years, except that there were now occasional convulsions. The patient died of pulmonary edema following a prolonged seizure.

**Post Mortem Findings:** At autopsy, an area of vascular malformation was found around the aqueduct of Sylvius, almost completely obstructing it (figs. 6 and 7). Hydrocephalus was moderate (fig. 8). There were many other small foci of vascular malformation. These were found elsewhere in the pons, in the cerebrum (fig. 8) and in the cerebellum. Histologically, the lesions in the cerebral hemispheres and pons were more compact than those of the other malformations described in this series (fig. 7). In the cerebellum, however, neuronal tissue was present among the abnormal vessels. There was considerable enlargement of blood vessels throughout the brain in regions outside the grossly visible foci. Otherwise, no tissue dysgenesis was detected. No extracranial vascular lesions were found.

In Case 5, there would seem to be an early disturbance of which the vascular malformation was easily visualized in gross. It is likely that changes in the anomalous vessels in adolescence produced some degree of hydrocephalus and that this progressed to almost complete blockage of the aqueduct. The moderate degree of hydrocephalus seen at the autopsy was not necessarily so great as that when the patient was in her thirties. Lange-Cosack (7) has reported similar findings in a group of patients found among a much larger number of subjects with psychotic states in whom arteriovenous angiomas of various types were demonstrated at autopsy.
It is a reasonable assumption that vascular development in all parts of the brain only occurs in response to the growing needs of the neural parenchyma. This would imply that any retardation in parenchymal development might have associated vascular irregularities. When such dysgenesis involves the brain stem and cerebellum, the leptomeningeal vessels are unusually numerous and large. Such "secondary" anomalies of vasculature include cavernous veins, large engorged capillaries, sinusoidal channels and smaller arterial type vessels with muscular walls. The large blood channels of both venous and arterial structure are increased in number while the actual capillary bed in association with neural parenchyma is reduced.

In a considerable number of patients with mental deficiency, present methods are insufficient to detect tissue changes which can be correlated with the clinical findings. In the brains of such patients, it is quite common to find large, even cavernous, blood channels in many parts of the brain, especially the cerebellum (fig. 9). These channels should not be regarded as a total picture of maldevelopment. For reasons given above, their presence is no indication of increased vascular supply to the neural parenchyma.
The range of clinical phenomena associated with very small vascular malformations is presented in a series of case studies. These include subarachnoid hemorrhage, focal seizures, intraventricular hemorrhage, hydrocephalus, and mental deficiency.

In some instances, the vascular malformations are a part of an area of tissue dysgenesis, involving neural parenchyma as well as vasculature. The small malformations resulting in subarachnoid and intraventricular hemorrhage are usually single lesions and those causing obstructive hydrocephalus may be.

Multiple small vascular anomalies are found in some patients with mental deficiency but are regarded as one part of a more complex process. Enlargement of the vascular tree in large areas of the brain without focal malformation is a common finding in patients with mental deficiency. These do not indicate increased vascular supply to the neural parenchyma.

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REFERENCES

DISCUSSION

Dr. G. A. Jervis, Thiells, New York: The speakers in these three papers have been so complete, that there is very little to add. My experience on hydrocephalus concerns some 50 pathological cases, most of them of the non-communicating type. The pathological findings were classified on a topographical basis following the scheme of Russell. The majority of cases were an example of what the speakers this afternoon called atresia of the aqueduct. Russell's term "forking" is probably to be preferred because it describes well the peculiar type of pathology. That "forking" is a manifestation of a dysraphic state is well known and is confirmed by the frequent occurrence of other dysraphic signs. Only in two cases of hydrocephalus was there an intensive gliosis of the periaqueduct without forking. Funnel-shaped canal was present above the stricture. This type is described often in the old literature. I would be inclined to think of these two cases as blastomatous gliosis, although differentiation between reactive and neoplastic glia is admittedly difficult.
Another curious finding in three cases was that of a pure stenosis without forking and without ependymal proliferation. I am well aware of the fact that similar stenosis without forking may be present in normal brains without hydrocephalus. In three other cases there was a thin septal formation blocking the aqueduct, one at the upper end of the aqueduct, and two at the level of the quadrigeminal bodies. In another exceptional case there was a small angioma occluding the lumen of the aqueduct.

The other type of hydrocephalus discussed by both Dr. Lichtenstein and Dr. Benda, occurring with Arnold-Chiari deformity was seen in some ten cases all accompanied by cervical meningocele. The condition has been well known for over fifty years, but the mechanism by which hydrocephalus develops is still not clear. Some fifteen years ago, Dr. Lichtenstein suggested that a traction upon the midbrain causes occlusion. Russell's theory that hydrocephalus arises from displacement of the fourth ventricle foramina into the spinal canal is more widely accepted. The new point in today's presentation was the description of cases of forking combined with Arnold-Chiari deformity, a finding as far as I know, not previously reported.

I would like to close, with a general observation. Discussion of anomalies of the nervous system may appear somewhat dry to the clinically oriented neurologist, yet there is a practical point in these studies which is frequently missed. There is much talk these days about research in the field of mental deficiency and cerebral palsy and much money is spent on it, with the ultimate hope of somehow cutting down the number of these unfortunate children by some kind of preventive medicine.

It is increasingly clear, I think, that a very large part of severe cases of mental deficiency and a substantial portion of patients with cerebral palsy are prenatally determined. In fact, major or minor malformations of the type presented here this afternoon, are frequently seen in this group of patients, and obviously are the results of pathogenetic factors operating during fetal life. If by morphologic and morphogenetic investigations we may pinpoint the period of the intrauterine life at which something went wrong, obviously a first and necessary step would be accomplished in our attempts to uncover and possibly to eliminate the responsible pathogenetic factors. Of course, quite a few malformations are controlled by genetic factors and, therefore, beyond our manipulations. However, clearcut, purely exogenous and controllable factors (such as some infections) are also known. In general the abnormality of development is the outcome of extremely complex interaction of genetic and non-genetic factors operating in mechnisms of growth and differentiation. A better understanding of the action of these factors presupposes an extensive knowledge of embryology and morphogenesis. Because of this, I think all of us whose work deals primarily with child neurology will be grateful to the authors for their excellent presentations.

Dr. K. T. Neuburger, Denver, Colorado: There is very little left for me to say after these thorough and informative presentations and after the discussion by Dr. Jervis, who has mentioned a number of things I had wanted to elaborate. I should like to emphasize, taking up where Dr. Jervis left off, that prenatal damages to the central nervous system play a much greater role in the encephalopathies under discussion than was assumed in former years, when practically everything was blamed on traumatic birth injury. Such damages include arrest of differentiation, prematurity, erythroblastosis, venous thrombosis, infections followed by hydrocephalus and others. Interference with oxygen supply is of prime importance, as recently emphasized by Towbin.

I shall restrict myself to a few words with regard to Dr. Lichtenstein's paper. I wish to stress the difficulty in distinguishing between dysplastic, reactive and neoplastic aqueductal stenosis. In some instances we have found it impossible to classify a given case without knowledge of the clinical picture.

In a recent paper on aqueductal occlusion, Colmant saw neoplasms in about one-half of his cases, especially juvenile protoplasmatic astrocytomas of the tegmentum, sometimes only pea-sized, poorly defined and usually composed of astrocytes, oligodendrocytes and microglia, with small cavities resembling central canals. We have seen such cases too.

Even severe aqueductal stenosis may be compatible with a relatively long duration of
life. A girl with hydrocephalus, idiocy and epilepsy since birth lived to be 16; autopsy showed periaqueductal gliosis, aqueductal stenosis and forking, microgyria, and a huge unilocular ventricular cavity with severe surrounding hemosiderosis. In the causation of the latter, head injury suffered in conjunction with multiple severe epileptic attacks may have played a part.

As far as traction stenosis is concerned, the gross picture resembles the vertically narrowed, oblong and curved aqueduct we sometimes see in slowly growing spongiosplastomas of the pons.

DR. W. J. GARDNER: With trepidation I arise to contribute to this symposium of pathologists, drawing encouragement from the fact that the neurological surgeon has the opportunity of observing the hydrodynamics of these pathologic states in the living patient.

I think that we are lumping together two forms of dysraphia. I am referring particularly to Dr. Benda's remarks. I believe that in addition to primary dysraphia, which represents failure or improper fusion of the neural tube, there is dysraphia secondary to hydrotension of the neural tube caused by hydrocephalus in the embryo.

There also is considerable evidence that the Arnold-Chiari malformation, like the Dandy-Walker syndrome (a term coined by Benda), is due to embryonal atresia of the outlets of the fourth ventricle; that the myelomeningocele of the former and the bulging occiput of the latter originate as diverticulae of the neural tube, produced in the embryo by the pressure of obstructive hydrocephalus; that the adult counterpart of myelomeningocele is syringomyelia.

Here is a slide showing the features which may be common to the adult and the infantile forms of the Dandy-Walker and Arnold-Chiari malformations. There is obstruction of the outlets of the fourth ventricle by membranes the attachments of which suggest that they are remnants of the embryonal rhombic roof. They are present in the adult and the infantile forms of both conditions. They are readily apparent in the Dandy-Walker deformity and in the adult form of the Arnold-Chiari malformation. They are difficult to demonstrate in the infantile form of the Arnold-Chiari malformation where they are obscured by the severity of the herniation or ruptured by the pressure of the hydrocephalus. There is hydrocephalus and hydromyelia in both the adult and infantile forms of both conditions. Increased intracranial pressure is not present in milder forms that go on to adulthood but it is severe in the infantile forms. The localized bulging of the cranio-vertebral axis is present only in the infantile forms where the pressure is severe. Basilar impression and scoliosis are found in the adult forms of both conditions.

Here is an operative photograph of the normal foramen of Magendie. One can see the cerebellar tonsils, the medulla and the obex of the fourth ventricle. After exposing this area at operation, the neurologic surgeon observes the cerebrospinal fluid spurting from the foramen of Magendie with each heart beat. The choroid plexus has been called a cerebrospinal fluid pump, and this is an apt term since, as a result of its pulsations, the fluid spurts from the foramen of Magendie.

Here is a diagram of the normal cerebrospinal spaces. With each pulsation of the choroid plexus, the fluid comes through the aqueduct and spurts out into the subarachnoid space. There the force of this ventricular fluid pulse wave is expanded on the outer surface of the spinal cord. This, we believe, is the reason that the central canal of the cord, which was originally connected to the fourth ventricle, closes off in the normal adult.

Now, if the foramen of Magendie fails to open, the central canal of the cord cannot close off and maintains its embryonal relationship as a part of the ventricular system. Throughout the rest of that individual's life, with each heart beat, the ventricular fluid pulse wave will be directed into the open end of the central canal causing it to gradually dilate. If the central canal ruptures, a diverticulum may form which will run up and down the posterior columns or horns to form a true syrinx parallel to the central canal.

This is an operative photograph of an adult form of Arnold-Chiari malformation. In 1950, we reported on 17 adult patients with this malformation verified at operation. Of these, 8 had scoliosis, 8 had basilar impression, and 13 had hydromyelia. In addition to the 13 who had proven hydromyelia, 2 others had dissociated sensory loss.
At operation for syringomyelia, the object should not be to incise the sac, but to establish the patency of the foramen of Magendie. In every patient with syringomyelia that we have operated upon, excepting only those cases due to tumor, we have found an obstruction of the foramen of Magendie.

The next slide will show the same case with the cerebellar tonsils separated by retractors. Here one can see the semiopaque membrane occluding the foramen of Magendie attached to the obex and cerebellar tonsils. When this membrane is opened, the ventricular fluid pulse wave escapes through the foramen of Magendie where it exerts its pressure on the outer surface of the cord as nature intended, with resulting reduction in size of the syrinx.

DR. F. WOHLWILL, Boston, Mass.: We have studied a case which belongs to Dr. Lichtenstein's last group. There was cervical spina bifida, Arnold-Chiari's syndrome and a complete atresia of the aqueduct. As in his cases, there was a very marked gliosis in the area of the remnants of the aqueductus. In my opinion, the problem of the relationship between the gliosis and the atresia has not been solved satisfactorily so far. It came to my mind that what Dr. Lichtenstein calls traction stenosis might be transformed into glial atresia. The fact that there is glial proliferation shows there is a process going on. It might be that the pathological conditions prevailing in Arnold-Chiari's syndrome provoke proliferation of the glia which, in turn, might interrupt the already stenotic aqueduct and then lead to a complete atresia. Otherwise the relationship between atresia and Arnold-Chiari's syndrome would be difficult to explain.

I was especially interested in Dr. Lichtenstein's case in which the dilated ventricles contained a coagulated substance with formation of a cellular wall. In our case there was a marked pyocephalus; the infection was probably due to leakage of an encephalomeningocele that existed at the same time and might have spread to the ventricles through the fissura transversa because there was no other way open for the infection. In our case the ventricles were completely surrounded by a pyogenic membrane consisting of fibroblasts, inflammatory elements and new-formed capillaries. It was so characteristic that when I saw the first sections which only contained part of the posterior horn, I diagnosed brain abscesses. Only afterwards I realized that I was dealing with pyocephalus which induced a reaction in the form of a pyogenic membrane. I believe that is a very rare event. Usually pyocephalus is lethal in a very few days probably because the infection reaches the 4th ventricle and the centers of the medulla oblongata are damaged; but as the aqueduct in this case was closed, there could be no infection of the 4th ventricle. In this way there was time for the development of the real pyogenic membrane. The child died at the age of forty days.

DR. B. J. ALPERS, Philadelphia, Pa.: It is customary to include in the arteriovenous anomalies of the nervous system the telangiectases, Sturge-Weber disease, arterial angiomata, venous angiomata, and arteriovenous aneurysms. There are a few features which all of these conditions have in common. The most striking is that the vascular disorder involves not only the surface, but it affects also the nervous tissue. Or as Bergstrand states it: Nervous tissue is found among the blood vessels in the entire group. Only for the telangiectases does there seem to be difference of opinion on this point. The nature of this nervous tissue is not neoplastic, though its arrangement may be disorderly and gliosis of varying degrees is found. Common to all also is a collection of blood vessels on the surface of the brain. In the case of the telangiectases and Sturge-Weber's disease, the entities appear to be relatively clearly defined, though in the case of the telangiectases the distinction from cavernous angiomata is not always possible. The problem is more difficult in regard to the other members of the group—the arterial and venous angiomas and the arteriovenous aneurysms. It is reasonable to ask whether all these may not in reality be members of a single group, all of them representing variations of arteriovenous aneurysms or anomalies. In this group the feeding arteries are hypertrophied and dilated; the draining veins dilated, pulsating and containing arterial blood; and the intervening tangle of blood vessels are for the most part undifferentiated but may have the structure of arteries or veins. The suggestion has been made that the arterial angiomas are in reality arteriovenous aneurysms, since the draining vessels have always proved to be veins rather than arteries. Similarly, as Cushing and Bailey announced many years ago, most venous angiomas, rare as they are, are
closely related to the arteriovenous aneurysms and in reported venous angiomas arterial blood is found. Bergstrand and Olivecrona believe the venous angiomas are indistinguishable from the arteriovenous aneurysms.

A study of both biopsy and autopsy specimens reveals the well-known fact that the visible arterial and venous enlargement on the surface of the brain represents only a small portion of the involved blood supply. Due to the manner of development of the anomalies the penetration of vessels from the surface and deep in the tissue produces a wedge-shaped conglomeration of vessels. In many instances most of the vessels in the anomaly are often hidden and the futility of ligation of the surface vessels is readily demonstrated by a study of removed specimens. It is not surprising, therefore, in view of the structure of these aneurysms, that hemorrhage may occur from the surface vessels and be subarachnoid or from the vessels within the parenchyma and be cerebral. While the majority of arteriovenous anomalies appear to be associated with the internal and external carotid arteries and their branches, they are found elsewhere and good examples are reported with involvement of the vein of Galen and the internal carotid arteries.

In the case of the arteriovenous aneurysms the developmental process appears to be the same, but whether it is the same for the other groups is hard to say. The defect lies apparently in an incomplete differentiation in the process of embryological development of the blood vessels, resulting in the occurrence of abnormal nests of blood vessels. The connection in very rare instances between the arterial and venous supply is direct. In the great majority of instances it consists of a tangle of blood vessels, surface and parenchymatous, with multiple arterial and venous channels. Little helpful information is obtained from a study of the blood vessels in the arteriovenous anomaly or aneurysm. The majority are abnormal and show various degrees of undifferentiation, and there is often lack of faulty development of one of the vessel coats, sometimes overgrowth of the intima, abnormal elastica.

The clinical correlations of the known pathology seem obvious. Mention has been made of the inadvisability of ligation of the surface blood vessels. It seems clear that if anything is to be done for these patients, complete removal of the arteriovenous anomaly is necessary, and indeed has frequently been carried out. The question arises whether such a removal effectively eliminates the possibility of recurrent bleeding. Postoperative arteriography in a few instances has revealed a reduction in the caliber and tortuosity of the feeding blood vessels, and a return to a normal vascular pattern in two to three weeks. There is one other feature of these anomalies which should be mentioned and that is the fact that the arterial supply in the arteriovenous anomalies is often multiple and concerns more than a single vessel.

DR. B. W. LICHTENSTEIN, Chicago, Illinois: In order to close the discussion of the first phase of our symposium I shall attempt to answer some of the questions. Dr. Gardner demonstrated lantern slides and discussed certain aspects of platybasia and the Arnold-Chiari complex. Since my presentation was specifically in regard to aqueductal stenosis and atresia in infants and young children his discussion was not exactly germane to the problems under consideration. I first discussed the problem of atresia of the aqueduct with neither spina bifida nor the Arnold-Chiari complex. Since my presentation was specifically in regard to aqueductal stenosis and atresia in infants and young children his discussion was not exactly germane to the problems under consideration. I first discussed the problem of atresia of the aqueduct with neither spina bifida nor the Arnold-Chiari complex. I then discussed 15 cases in which obstruction was in the nature of atresia identical with the changes seen in the first part of my presentation. In the remaining nine cases in which the aqueductal obstruction was associated with spina bifida, hydrocephalus and the Arnold-Chiari complex, the aqueduct did not exhibit the features of atresia but was elongated, narrow, and slit-like. This is the prime object of my presentation. Inspection of the pons revealed it to be elongated, narrowed, with a deep groove cut by the basilar artery. In short, the narrowing of the aqueduct and its obstruction without atresia appeared to be due to traction.

The term Arnold-Chiari complex has been used to designate more than one pathologic process. If the term is defined as an alteration of the hindbrain and the localization of cerebellar tissue and portion of the medulla oblongata below the foramen magnum, then we
must recognize at least two major varieties. The first in association with spina bifida and which probably is due to downward traction and the second in association with platybasia and other bony anomalies and deformities in which the relationship of the neuraxis to the skeletal axis is altered and hind-brain tissues descend into the upper cervical spinal canal. In the latter instances the aqueduct is not elongated and the pons is not deformed as described above.

I realize that even for the simple atresia of the aqueduct there is no consensus since, as Dr. Neubürgger emphasized, the differentiation of blastomatosis gliosis from reactive and malformative gliosis may be very difficult.