

ANATOMIC FEATURES COMMON TO THE ARNOLD-CHIARI AND THE DANDY-WALKER MALFORMATIONS SUGGEST A COMMON ORIGIN

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. . . . in every art, fundamental matters are
perennially being discovered, discredited, forgotten,
rediscovered and reaffirmed

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MILD examples of the Arnold-Chiari and Dandy-Walker malformations were described in adults by Chiari,^{2,3} by Walker,⁴ as well as by subsequent authors. Gardner, Abdullah, and McCormack⁵ have shown that the mild examples of these two malformations found in adults have so many anatomic features in common that they must arise from a common cause, namely, embryonal atresia of the fourth ventricle. The purpose of this communication is to point out that the same is true of the severe forms encountered in the hydrocephalic infant.

Definition of Terms

Chiari,² in 1891, described in detail three degrees of a deformity of the hind-brain that he attributed to hydrocephalus of the forebrain. This description appeared three years prior to Arnold's⁶ incidental and quite incomplete account of the same deformity. Chiari's type 1 deformity, now usually referred to as a "pressure cone," consisted essentially of a herniation of the cerebellar tonsils through the foramen magnum. His type 2, subsequently entitled the "Arnold-Chiari malformation" by Arnold's students,⁷ consisted of a herniation of the inferior vermis, pons, medulla, and the compressed fourth ventricle through the foramen magnum with a steplike formation of the medulla and a shortening of the cervical cord. In his type 3, the cerebellum was herniated into a high cervical meningocele. In his commonly quoted second paper, Chiari³ reported 14 cases of type 1, all in adults or adolescents, only one of whom had myelomeningocele, and seven cases of type 2, all in infants with myelomeningocele. He pointed out that both types were caused by hydrocephalus and that type 2 was merely a severe form of type 1. Some authors since Chiari have denied that his type 1 (pressure coning) is related to his type 2 (Arnold-Chiari malformation) and, in the latter condition, have substituted cause for effect by attributing the hydrocephalus to the malformation.

It is unfortunate that the term "Arnold-Chiari malformation" has become so firmly established, because this term confuses the issue by referring only to Chiari's type 2 and thus tends to set it apart from Chiari's type 1. My observations support

Chiari's convictions that type 1 is a mild form of type 2 and that both are the result of hydrocephalus, and I therefore shall revert to Chiari's terminology.

The term "Dandy-Walker syndrome" was introduced by Benda⁸ to describe a different deformity of the hindbrain in which, instead of compression, there is dilatation of the fourth ventricle. This deformity, also caused by hydrocephalus, was attributed by Dandy⁹ to congenital occlusion of the foramen of Magendie and Luschka. This occlusion subsequently was shown by Taggart and Walker¹⁰ to result from lack of perforation of the embryonic rhombic roof. In this malformation, as in Chiari's, severe forms usually are discovered in infants and mild forms in adolescents or adults. In the Dandy-Walker deformity the posterior attachment of the tentorium is higher than normal, and the hydrocephalus, therefore, affects the hindbrain more than the forebrain (*Figs. 1-3*). In the Arnold-Chiari malformation, hereinafter referred to as the Chiari type-2 malformation, the attachment of the tentorium is lower than normal and, since this is accompanied by compression of the hindbrain and frequently also of the midbrain, the hydrocephalus can affect only the forebrain (*Fig. 4*).

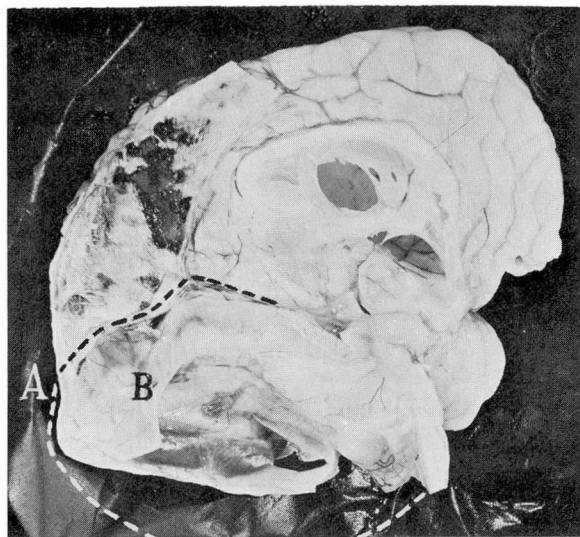


Fig. 1. Dandy-Walker malformation in a hydrocephalic infant seven and one-half months old. The sagittally sectioned forebrain and hindbrain have been reassembled. The dotted line represents the approximate limits of the enormously distended posterior fossa. The junction of the sagittal and lateral sinuses is at A. The membrane seen to the left of the attenuated cerebellum is the left leaf of the diaphragm. It includes the falx cerebelli B and is lined by the transparent roof of this enormous fourth ventricle. Note also the flattening of the brain stem. This specimen was hardened in situ by embalming the body and injecting concentrated formalin into the ventricles six hours prior to necropsy.

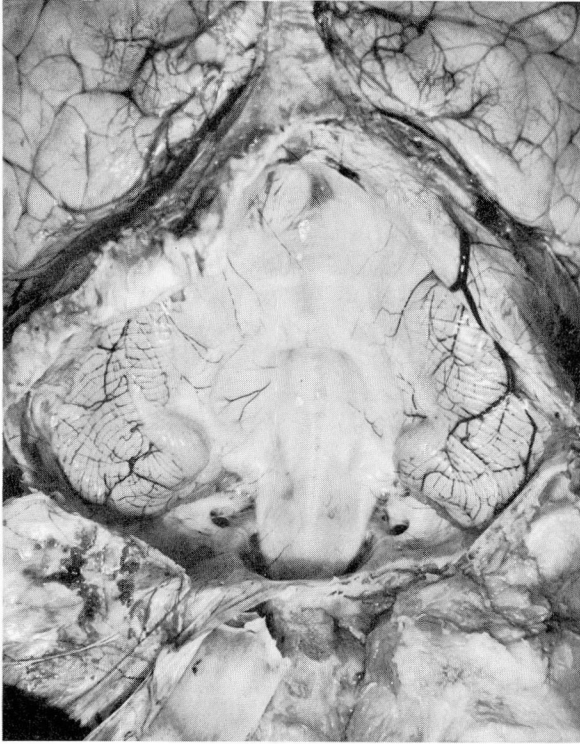


Fig. 2. Dandy-Walker malformation, the same case as that of *Figure 1*. Posterior aspect of the brain before removal from the skull. The great depth of the cavity is not apparent in this photograph. The junction of the sagittal with the lateral sinuses above the level of the lambdoid sutures forms a Y rather than a T. The tentorium, the small cerebellar lobes and the attenuated vermis are dislocated cephalad. The brain stem is flattened, widened, and elongated so that the floor of the fourth ventricle extends through the foramen magnum. In most of the reported cases only the membranous roof has been herniated through the foramen. The jugular foramina stand out prominently. Above and lateral to them may be seen strands of choroid plexus.

Anatomic Features

It is a medical axiom that to develop a proper concept of the nature of a disease process, one should begin with a study of mild cases. Anatomic features shared by the mild (usually adult) cases of Chiari and Dandy-Walker malformations are as follows:⁵ compensated hydrocephalus, either communicating or noncommunicating; hydromyelia (dilatation of the central canal); syringomyelia (syrinx or diverticulum communicating with the central canal); basilar impression; scoliosis; thickening of the meninges at the level of the foramen magnum; and closure of the foramina of the fourth ventricle by membranes that represent persisting portions of the embryonic rhombic roof.

Anatomic features common to the severe (usually infantile) forms of these two malformations will be shown to be as follows: uncompensated hydrocephalus, either communicating or noncommunicating; hydromyelia; syringomyelia, myelomeningocele; encephalocele; thickening of the meninges at the level of the foramen magnum; dislocation of the caudal portion of the fourth ventricle into the spinal canal and closure of the foramina of the fourth ventricle by membranes that represent persisting portions of the embryonic rhombic roof.

Hydrocephalus. The progressive enlargement of the head which occurs in most infants with the Chiari type-2 malformation, indicates that the hydrocephalus is uncompensated. Russell and Donald¹¹ showed that this hydrocephalus may be either communicating or noncommunicating.

In the Dandy-Walker malformation of infancy, the accompanying hydrocephalus also is uncompensated. In three cases described by Benda,⁸ necropsy disclosed that one was noncommunicating and two were communicating. In four cases reported by Schreiber and Reye,¹² antemortem tests indicated that the hydrocephalus was noncommunicating in one and communicating in three. In Matson's¹³ surgical experience with eight cases of this condition, opening the occluded fourth ventricle relieved the hydrocephalus in three cases but in the

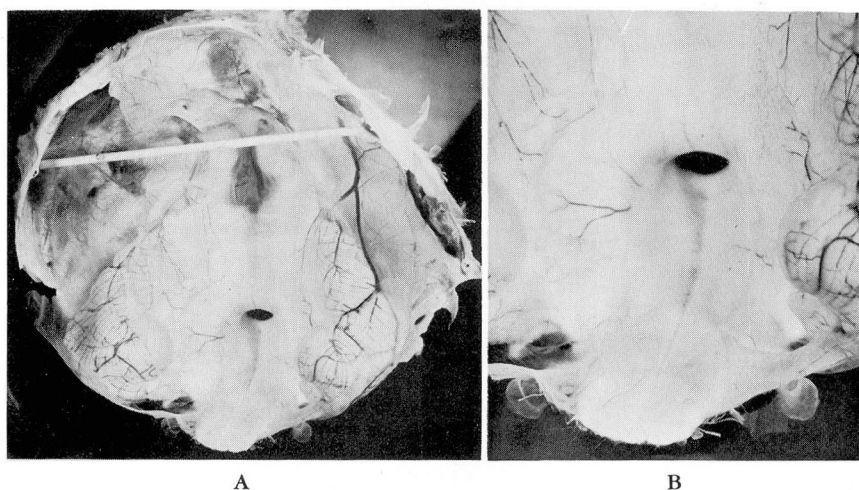


Fig. 3. Dandy-Walker malformation, the same case as that of *Figures 1 and 2*. A, The hindbrain and attached tentorium have been removed and floated in water. One can see the torn edge of the delicate membrane (rhombic roof) that previously had formed an invisible lining of the entire inner surface of the dura and the tentorium and had herniated through the foramen magnum. It is attached to the taenia and to the cerebellum at its line of contact with the dura and the tentorium. On either side of the transected medulla, a tiny outpouching of the delicate membrane may be seen which had herniated into the jugular foramina. B, These are better shown in the enlargement. Similar outpouchings into the acoustic meatuses apparently had been divided along with the eighth cranial nerves.

remaining five it served merely to "convert" the noncommunicating hydrocephalus into the communicating variety. Therefore, in the Dandy-Walker as in the Chiari type-2 malformation, the uncompensated hydrocephalus may be either communicating or noncommunicating, or a combination of both, with inadequate absorptive capacity of the subarachnoid space superimposed upon partial ventricular obstruction. Ventricular obstruction in no case can be really complete because all living tissues, including the walls of the ventricles, are permeable to water in some degree.

Hydromyelia and syringomyelia. The infant with a Chiari type-2 malformation usually has hydromyelic dilatation of the central canal of the cord. This relationship, first described by Chiari,^{2,3} was demonstrated by Ingraham and Scott¹⁴ in 8 of 20

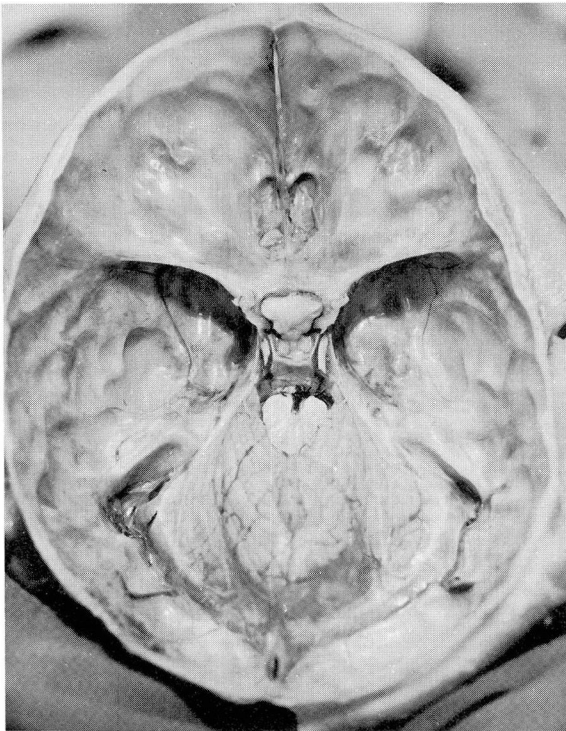


Fig. 4. A case of Chiari type-2 malformation in a child aged four years and five months with congenital hydrocephalus and myelomeningocele. The same method of fixation was employed as described in Figure 1. The hydrocephalic forebrain has been removed. The entire attachment of the tentorium is dislocated caudally including the superior petrosal sinuses which are depressed well below the petrous ridges. Note that the cerebral peduncles are displaced downward behind the clivus into the posterior fossa while the superior surface of the vermis bulges upward through the incisura. The aqueduct, compressed in the midbrain hernia, is not visualized. This necropsy study illustrates the advantages of fixation in situ and removal of the hydrocephalic cerebral hemispheres before disturbing the relationship of the structures of the posterior fossa.

cases, and by Cameron¹⁵ in 20 of 22 cases. An infant with this malformation, also may have a true syrinx or cavitation paralleling the dilated central canal of the cord as was described by Lichtenstein¹⁶ (Fig. 5) and by Cameron.¹⁵

Benda⁸ found the combination of hydromyelia and syringomyelia also in the infant with Dandy-Walker malformation. In his case 3 he described "congenital syringomyelia of the hydromyelic type" throughout the entire length of the spinal cord. The gray matter of the cord had failed to differentiate in the normal way, and the central canal formed a large slit walled by ependymal cells. At some levels the cavity was enlarged and filled with spinal fluid while at others a secondary cavity was formed within the gray matter. In his case 4B the gray matter of the cord "was completely undifferentiated, forming a central syringomyelia." Thus it is apparent that the infantile forms of the Dandy-Walker and of the Chiari malformations may be accompanied by hydromyelia and a syrinx as well.

Inasmuch as the ventricles and the central canal of the cord constitute a single cavity, the term "hydrocephalomyelia" has been proposed for the state in which both are dilated.¹⁷

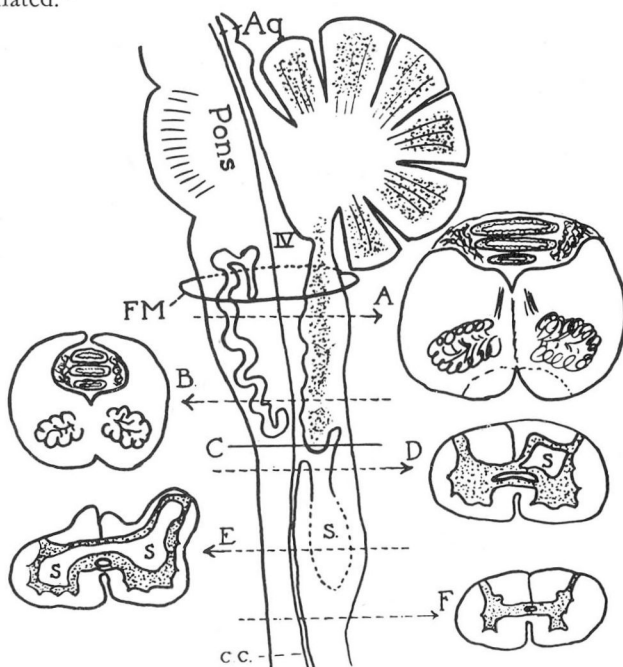


Fig. 5. Schematic drawing of a midsagittal section through the hindbrain and upper cervical cord of an infant with hydrocephalus, Arnold-Chiari malformation, and myelomeningocele. FM indicates the foramen magnum, S the syrinx, cc. the central canal, and C the level of transection at necropsy. (Courtesy of Lichtenstein, B. W.: *Arch. Neurol. & Psychiat.* 49: 881-894, p. 890, June 1943; A.M.A. *Arch. Neurol. & Psychiat.*) Note the lack of perforation of the rhombic roof (embryonic atresia of the fourth ventricle).

Myelomeningocele. The association of the Chiari type-2 malformation with myelomeningocele described by Chiari,² recently has been restudied by Cameron¹⁵ who, in 26 infants with myelomeningocele, found the Chiari type-2 malformation in its typical form in 22, "atypical variants" of it in two (one of these was Chiari's type 1), and no malformation in the other two.

It is not generally recognized that an infant with Dandy-Walker malformation also may have myelomeningocele. Dandy⁹ shows a photograph of such a patient, and Benda⁸ describes a myelomeningocele in his case 3 referred to above. Myelomeningocele, therefore, is still another feature associated with the severe Dandy-Walker as well as with the Chiari type-2 malformation.

Encephalocele. My survey of the literature to date has disclosed no fully documented cases of encephalocele associated with Chiari type-2 malformation unless it be those of Cameron.¹⁵ In this author's¹⁵ 26 cases of "spina bifida" in infants, there were two cases in which there was coexisting "cranium bifidum." The roentgenogram of one case showed a large encephalocele and severe craniofacial anomaly. This latter condition Cameron¹⁵ found only in severe cases of Chiari type-2 malformation, so the reader must assume that this patient had both encephalocele and Chiari type-2 malformation although the author does not specifically so state.

The association of encephalocele with the Dandy-Walker malformation, on the other hand, is well documented. Bland Sutton¹⁸ in 1887, described and illustrated a case in which the cerebellum was "virtually absent" and a large encephalocele containing choroid plexus protruded from a bulging posterior fossa. Dandy¹⁹ also showed an illustration of an encephalocele protruding through a hiatus in the occipital bone, communicating with a greatly distended fourth ventricle, and "participating in the hydrocephalus caused by a block of the foramen of Magendie" (Fig. 6). Schreiber and Reye¹² described two cases of occipital encephalocele communicating with a Dandy-Walker malformation of the posterior fossa. Encephalocele, therefore, is encountered in the infant with the Dandy-Walker and perhaps also with the Chiari type-2 malformation.

Membranes closing the foramina of the fourth ventricle. Taggart and Walker¹⁰ described closure of the enlarged foramina of the fourth ventricle by persisting portions of the membranous rhombic roof as the cause of their "congenital atresia of the foramina of Magendie and Luschka" (Benda's⁸ Dandy-Walker syndrome). Subsequent authors, with one exception (Benda⁸), have accepted this explanation. Histologically similar though less expanded membranes occluding the foramina of Magendie and Luschka may be readily demonstrable in the Chiari type-1 malformation. Russell's²⁰ case 19 is an example (Fig. 7). In this infant, whose spinal cord was tethered at the level of a lumbosacral meningocele, she described the brain as follows: "There was gross internal hydrocephalus, involving all ventricles; the aqueduct measured 0.5 cm. in diameter. At the foramen magnum the medulla was closely enveloped by a cone-shaped mass of cerebellar tissue . . . which proved,

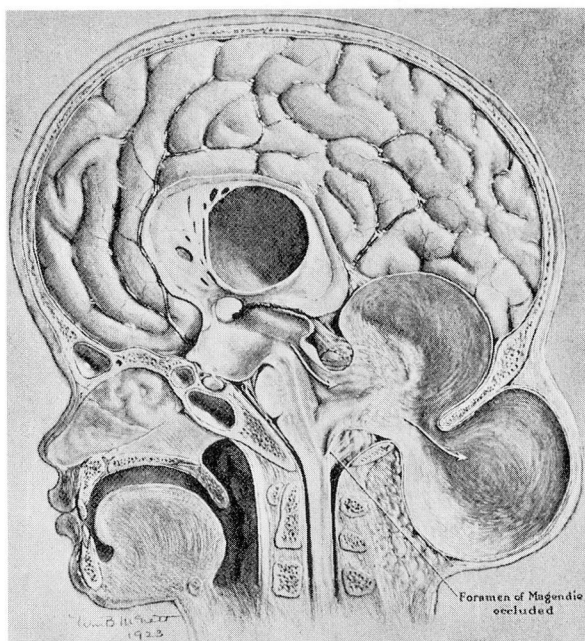


Fig. 6. Dandy-Walker malformation with encephalocele. (Courtesy of W. E. Prior Co., Inc.; Dandy, W. E.: *The Brain*. In Lewis, D. D.: *Practice of Surgery*, 1932, p. 258.

on dissection, to be composed of the tonsils. When separated, a translucent membrane was found at the site of the foramen of Magendie . . . There was great ballooning of the tissue at the foramina of Luschka, forming a translucent bleb, about 1 cm. in diameter, on either side . . . In a mid-sagittal section the membrane occluding the foramen of Magendie is composed, above and below, of fibrillary neuroglia lined, towards the ventricle, with a layer of ependymal cells. The central third of the structure is occupied by a flattened mass of choroid plexus. Externally the membrane is coated with the pia, which is reflected on to its surface from the adjacent cerebellum and medulla oblongata.” Russell,²⁰ in this case, is describing a Chiari type-1 malformation in association with what many authorities would consider an abortive myelomeningocele. Chiari,³ and Cameron¹⁵ as well, has described myelomeningocele in association with a Chiari type-1 malformation. Russell’s²⁰ histologic description of this membrane occluding the foramen of Magendie corresponds closely with that of her case 15 which she considered a probable example of the Dandy-Walker malformation.

Dandy may have been the first to recognize that the Chiari type-2 malformation is the result of obstruction of the foramina of Magendie and Luschka. In a paper⁹ on hydrocephalus due to occlusion of these foramina, he includes the following case: “In a fourth case, a dorsal meningocele was present at birth and



Fig. 7. Membrane obstructing the foramen of Magendie in an infant with a Chiari type-1 malformation, a lumbosacral meningocele, and a tethered cord. Each foramen of Luschka also was closed by a bulging membrane in this case. (Courtesy of Russell, D. S.: M. Res. Council Special Rep. Ser. No. 265, p. 34; H. M. Stationery Office, 1949; the Controller, H. M. Stationery Office, 1959.)

hydrocephalus was observed 2 months after birth. A dense scar covered the foramen of Magendie and the contiguous region. The cerebellum was firmly bound to the medulla and pons, there being no bulging fourth ventricle between. A large cerebellar hernia projected into the spinal canal. The pia-arachnoid was adherent to the dura over most of the area exposed at operation." Dandy recognized, despite the lack of dilatation of the fourth ventricle in this case, that obstruction of the foramens of Magendie and Luschka was the cause of the hydrocephalus and of the hernia of the hindbrain. He attributed the associated thickening and adhesions of the leptomeninges to inflammation. Others have reached a similar conclusion even in cases where microscopic study has revealed the presence of neuroglial elements and little or no evidence of inflammation. In the Chiari type-2 malformations the foramens of Luschka are so compressed in the spinal canal that they are almost impossible to study, but a membrane occluding the foramen of Magendie may be demonstrated in some cases (*Figs. 8 and 9*).

Thus, it is apparent that in the Chiari type-1 and type-2 malformations, as in the mild and the severe Dandy-Walker malformations, the foramens of the fourth ventricle may be occluded by membranes* that, in view of their attachments, obviously represent persisting portions of the embryonic rhombic roof. That these delicate membranes are not always found is not surprising in view of the

*Not yet proved for the foramens of Luschka in the Chiari type-2 malformation.

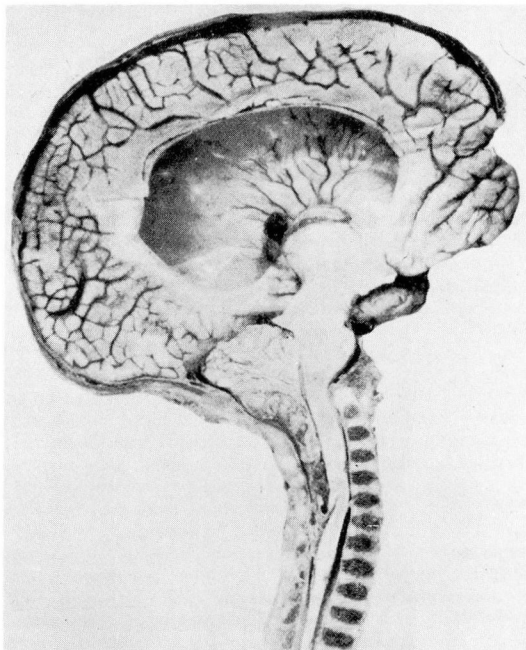


Fig. 8. Chiari type-2 malformation in an infant. (Courtesy of Moncrieff, A., and Norman, R. M.: Greenfield, J. G.: *Neuropathology*: Edward Arnold (Publishers) Ltd., 1958, p. 314.) The attachment of the bulging membrane to the cerebellar hernia and the brain stem indicates that it is the rhombic roof. Compare with *Figures 5, 7, and 9*.

severe stresses imposed upon them by the squeezing of the aftercoming hydrocephalic head in its passage through the birth canal.

Thickening and fusion of the leptomeninges at the level of the foramen magnum often is present in the Chiari type-1 and type-2 malformations. It has also been described in both the mild and the severe forms of the Dandy-Walker malformation, as in case 1 of Gardner, Abdullah, and McCormack⁵ and in case 1 of Taggart and Walker.¹⁰

The occluded lower end of the fourth ventricle is herniated through the foramen magnum both in the severe Dandy-Walker (*Fig. 2*) and the Chiari type-2 malformations (*Figs. 5, 8, and 9*).

Discussion

The neural tube of the embryo is a closed cavity consisting of the primitive ventricles and the central canal of the cord. Its lumen is large compared to the thickness of its walls, a relationship which in later life constitutes hydrocephalus and hydromyelia (hydrocephalomyelia). This disproportion of lumen to neural tissue, is a feature of normal embryonal development and, as shown in Weed's²¹

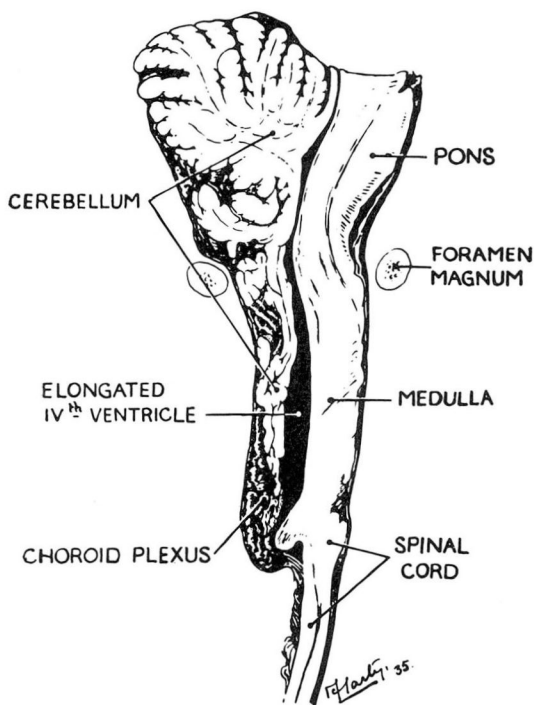


Fig. 9. Chiari type-2 malformation. (Courtesy of Russell, D. S.: M. Res. Council Special Rep. Ser. No. 265; H. H. Stationery Office, 1949, p. 23; the Controller, H. M. Stationery Office, 1959.) As in *Figure 8*, the cerebellar hernia appears to have pulled the attached brain stem down with it as evidenced by the posterior eversion of the obex and the resultant kinking of the upper end of the central canal.

illustrations, it reaches its most advanced degree shortly after the choroid plexus begins to elaborate fluid. At this stage the distended head of the embryo is a thin-walled, almost transparent cyst (*Fig. 10*). The normal embryo recovers from this "physiologic hydrocephalus" as it becomes compensated by the increasing permeability of the rhombic roof and of the absorbing areas in the subarachnoid space.

Weed²¹ believed that, if the rhombic roof were not sufficiently permeable at this critical period, the retained fluid would overdilate the lumen of the neural tube. In this case it would not filter through the roof in a quantity sufficient to accomplish adequately the job of dissecting open the developing subarachnoid spaces; that if this happened, the obstructive hydrocephalus would result in communicating hydrocephalus. This significant observation made in 1917 has been largely forgotten in present-day attempts to explain the origin of congenital hydrocephalus.

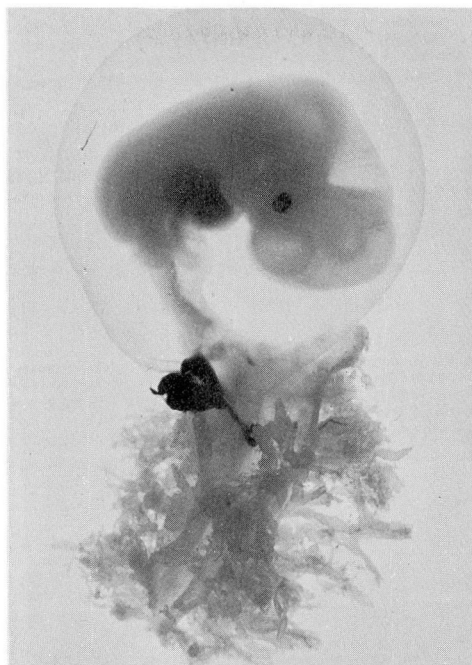


Fig. 10. At six weeks, when the choroid plexus begins to function,²¹ the head of the human embryo is a thin-walled bag of water. The most translucent area is the rhombic roof. This specimen was photographed after it had been made relatively opaque by formalin fixation.

The pulsating choroid plexus, a relatively enormous structure in the embryo acts like an unvalved pump to generate a pulse wave in the ventricular fluid.²² As long as the rhombic roof does not perforate, this pulse wave is funneled into the relatively large central canal. After the roof perforates to form the foramen, the ventricular fluid pulse wave is shunted into the spinal subarachnoid space. Thereafter the bypassed central canal, containing no choroid plexus and subjected to this pulsating pressure on its outer surface, narrows and becomes a vestigial structure. However, it will dilate again, in the experimental animal, if the foramen of the fourth ventricle are occluded by the reaction resulting from the intracisternal injection of kaolin.²³

Hydrocephalomyelia is present prior to the opening of the foramen of the fourth ventricle in the normal embryo; it is also present in the infant with Chiari type-2 malformation, and in the infant with Dandy-Walker malformation.* This implies that in these newborn infants, this state, which is normal only in the embryo, has been preserved because of a failure of the foramen of the fourth ventricle to open (embryonal atresia). The anatomic attachment and the microscopic appearance

*Hydrocephalomyelia also is present in the cases of mild Chiari and Dandy-Walker malformations in adults.^{5,24}

of the membranes that enclose the foramina in these malformations support this concept.

The hydrodynamic stresses responsible for these malformations may be compared to the stresses existing when the inflating pressure in an automobile inner tube exceeds the elastic resistance of the weakest portion of its wall; as this occurs, and despite the fact that the pressures in all parts of the tube are identical, a localized bulging and attenuation develops *with a consequent pushing away of the surrounding zone*. Transferred to the embryo with inadequate permeability of the rhombic roof, this phenomenon may affect the covering of either the hindbrain or the forebrain. If, in a given case, the covering of the hindbrain happens to offer less resistance, it will expand disproportionately and push the tentorium and the forebrain in a cephalad direction (Dandy-Walker malformation). On the other hand, if the covering of the forebrain happens to be more yielding, it will expand and displace the tentorium and the compressed hindbrain in a caudad direction (Chiari type-2 malformation). In the latter case the herniation of the hindbrain structures through the foramen magnum will meet relatively little resistance from the immature dentate ligaments. The herniation will be exaggerated in those cases in which the caudal portion of the neural tube is open (myelomeningocele).

If the permeability of the rhombic roof is only mildly or temporarily impaired, there may be little dislocation of the tentorium and the hydrocephalus may result in a mild Dandy-Walker or in a Chiari type-1 malformation. Such a mild degree of embryonal hydrocephalus may become compensated so early in intrauterine life that the ventricles are restored to normal size by the time of birth. However, in these cases, the central canal may maintain its patency as an offshoot of the closed ventricular system. The pulsations of the ventricular fluid then are funneled into it throughout life, and slowly progressing dilatation of the central canal may result in the clinical picture of syringomyelia.⁵ In the Chiari type-2 malformation, the upper end of the central canal may be doubled back on itself and closed off in the posterior buckling of the medulla (*Figs. 8 and 9*). This buckling is due to the fact that the rhombic roof is more free to move caudally than is the floor which must push the cervical cord ahead of itself.

The neurologic surgeon is accustomed to the concept that dilatation occurs uniformly in all portions of the ventricular system anterior to a point of obstruction.* This concept has made it reasonable for him to believe that the Dandy-Walker and Chiari type-1 malformations are due to obstruction of the outlets of the fourth ventricle.† However, this same concept has obscured his thinking in regard to the

*This concept applies to the brain in the nonexpandable skull where the ventricular dilatation is due to uniform compression of brain tissue; it does not apply to the brain within the soft, immature skull in which one part will expand more than another with resulting unequal stretching and attenuation of the underlying brain.

†The exaggerated dilatation of the fourth ventricle in the Dandy-Walker syndrome cannot be the result of a higher pressure in this ventricle since it communicates freely with the other ventricles. The unequal dilatation, therefore, can be explained only on the basis of a difference in the yielding qualities of the coverings.

Chiari type-2 malformation in which the downward displacement of the tentorium has compressed and narrowed the lumen not only of the hindbrain (fourth ventricle) but also, in some instances, the lumen of the midbrain with resulting stenosis and forking (due to longitudinal wrinkling of the wall) of the aqueduct. Furthermore, the stretching of the midline structures in the sagittal plane, which results from the increasing *sagittal diameter* of the expanding skull, may cause an approximation of the lateral walls of the third ventricle with thickening (Cameron¹⁵) rather than attenuation of the massa intermedia (Fig. 11). This stretching of the midline structures in the sagittal plane also may be responsible for attenuation of the falx (Cameron¹⁵), for fenestration of the septum pellucidum, and for dilation of the foramens of Monro.

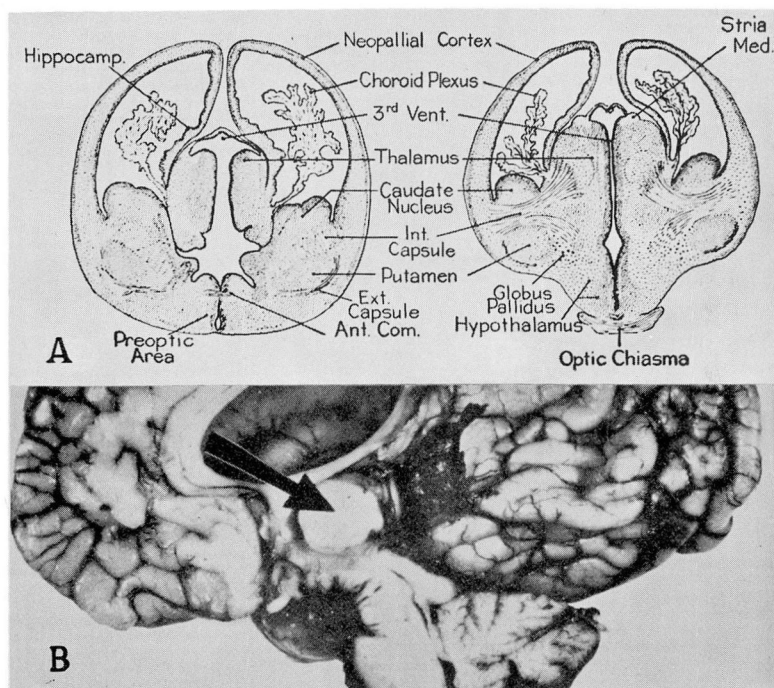


Fig. 11. A, Coronal sections of the brain of a three-month-old fetus. (Courtesy of Patten, B. M.: *Human Embryology*, Ed. 2; modified after Kodama; McGraw-Hill Book Co., Inc., 1953, p. 353.) It is apparent at this stage that ballooning of the membranous skull would result in stretching of the structures in the sagittal plane, which would result in closer approximation of the thalami and widening of their area of contact, i. e., of the massa intermedia. B, Brain of an infant with Chiari type-2 malformation. The arrow points to the interthalamic ramus, greatly thickened by approximation of the thalami.¹⁵ (Courtesy of Cameron, A. H.: *J. Path. & Bact.* 73: 195-211, Jan. 1957.) Note also the deformity of the quadrigeminal plate ascribed by Feigin²⁸ to developmental arrest, but more likely the result of downward dislocation and compression of the midbrain.

It is planned, in a future communication, to "rediscover," and perhaps to "re-affirm," Morgagni's²⁵ belief, which was later "discredited" by Von Recklinghausen²⁶ and has since been all but "forgotten," that myelomeningocele results from the disruption of a neural tube that has closed rather than from a failure of the tube to close.

Conclusions

The several anatomic features common to the Chiari type-2 and to the Dandy-Walker malformations suggest: that these deformities must have a common cause; that Chiari's hindbrain malformation is the result of hydrocephalus as he² stated in 1891, and not the cause of it as was advocated by Russell and Donald¹¹ in 1935, nor is the deformity produced by traction of the tethered cord as suggested by Penfield and Coburn²⁷ in 1938; that Chiari's type-1 malformation (pressure coning) is, as he^{2,3} stated, a mild form of his type-2 (Arnold-Chiari) malformation; that, as proposed by Weed²¹ in 1917, inadequate permeability of the embryonic rhombic roof may cause congenital hydrocephalus, both obstructive and communicating; that the Chiari type-2 and Dandy-Walker malformations with their accompanying hydrocephalomyelia are due to embryonal atresia of the fourth ventricle; that a normal hindbrain results if in embryonic life the rhombic roof differentiates properly and opportunely so that it is adequately permeable when the choroid plexus begins to function; that if the roof becomes adequately permeable a bit later than it normally should a Chiari type-1 or mild Dandy-Walker malformation may result; that if it becomes adequately permeable much too late or not at all a Chiari type-2 or a severe Dandy-Walker malformation may result.

Acknowledgment

I should like to acknowledge my indebtedness and, at the same time, to extend my apologies to Dr. Dorothy S. Russell, to Dr. B. W. Lichtenstein and to Dr. Clemens Benda for making such free use of their material, and to Dr. A. H. Cameron, whose careful, painstaking study of 26 cases of spina bifida disclosed hitherto-undescribed anomalies associated with the Chiari type-2 malformation and which also stressed the frequency of other significant features that have led me to differ with him regarding the nature of the hydrodynamic stresses that cause them.

References

1. Homans, J. (Comp.): *A Textbook of Surgery*, Ed. 4, p. 34. Springfield, Ill.: Charles C Thomas, Pub., 1936, 1267 pp.
2. Chiari, H.: Ueber Veränderungen des Kleinhirns, des Pons und der Medulla oblongata in Folge von congenitaler Hydrocephalie des Grosshirns. *Deutsche med. Wchnschr.* 27: 1172-1175, 1891.
3. Chiari, H.: Ueber Veränderungen des Kleinhirns, des Pons und der Medulla oblongata in Folge von congenitaler Hydrocephalie des Grosshirns. *Denkschr. d. k. Akad. d. Wissensch. Math.-naturw. Kl.* 63: 71-116, 1895.

4. Walker, A. E.: Case of congenital atresia of foramina of Luschka and Magendie; surgical cure. *J. Neuropath. & Neurol.* 3: 368-373, 1944.
5. Gardner, W. J.; Abdullah, A. F., and McCormack, L. J.: Varying expressions of embryonal atresia of fourth ventricle in adults. Arnold-Chiari malformation, Dandy-Walker syndrome, "arachnoid" cyst of cerebellum, and syringomyelia. *J. Neurosurg.* 14: 591-607, 1957.
6. Arnold, J.: Myelocyste, Transposition von Gewebskeimen und Sympodie. *Beitr. path. Anat.* 16: 1-28, 1894.
7. Schwalbe, E., and Gredig, M.: Über Entwicklungsstörungen des Kleinhirns, Hirnstamms und Halsmarks bei Spina bifida. *Beitr. path. Anat.* 40: 133-194, 1907.
8. Benda, C. E.: *Developmental Disorders of Mentation and Cerebral Palsies*. New York: Grune & Stratton, Inc., 1952, 565 pp.
9. Dandy, W. E.: Diagnosis and treatment of hydrocephalus due to occlusions of foramina of Magendie and Luschka. *Surg. Gynec. & Obst.* 32: 112-124, 1921.
10. Taggart, J. K., Jr., and Walker, A. E.: Congenital atresia of foramina of Luschka and Magendie. *Arch. Neurol. & Psychiat.* 48: 583-612, 1942.
11. Russell, D. S., and Donald, C.: Mechanism of internal hydrocephalus in spina bifida. *Brain* 58: 203-215, 1935.
12. Schreiber, M. S., and Reye, R. D. K.: Posterior fossa cysts due to congenital atresia of foramina of Luschka and Magendie. *M. J. Australia* 2: 743-748, 1954.
13. Matson, D. D.: Prenatal obstruction of fourth ventricle. *Am. J. Roentgenol.* 76: 499-506, 1956.
14. Ingraham, F. D., and Scott, H. W., Jr.: Spina bifida and cranium bifidum; Arnold-Chiari malformation; study of 20 cases. *New England J. Med.* 229: 108-114, 1943.
15. Cameron, A. H.: Arnold-Chiari and other neuro-anatomical malformations associated with spina bifida. *J. Path. & Bact.* 73: 195-211, 1957.
16. Lichtenstein, B. W.: Cervical syringomyelia and syringomyelia-like states associated with Arnold-Chiari deformity and platybasia. *Arch. Neurol. & Psychiat.* 49: 881-894, 1943.
17. Gardner, W. J., and Goodall, R. J.: Surgical treatment of Arnold-Chiari malformation in adults. Explanation of its mechanism and importance of encephalography in diagnosis. *J. Neurosurg.* 7: 199-206, 1950.
18. Bland Sutton, J.: Lateral recesses of fourth ventricle; their relation to certain cysts and tumours of cerebellum, and to occipital meningocele. *Brain* 9: 352-361, 1886.
19. Dandy, W. E.: The Brain. In Lewis, D. D.: *Practice of Surgery*, vol. 12, p. 258. Hagerstown: W. E. Prior Co., Inc., 1932.
20. Russell, D. S.: Observations on the Pathology of Hydrocephalus. Medical Research Council Special Report Series No. 265, pp. 32-33. London: His Majesty's Stationery Office, 1949, 138 pp.
21. Weed, L. H.: The Development of the Cerebro-Spinal Spaces in Pig and in Man. Washington, D.C.: Carnegie Inst., 1917, 116 pp., 4°.
22. Bering, E. A., Jr.: Choroid plexus and arterial pulsation of cerebrospinal fluid; demonstration of choroid plexuses as cerebrospinal fluid pump. *A.M.A. Arch. Neurol. & Psychiat.* 73: 165-172, 1955.

23. McLaurin, R. L.; Bailey, O. T.; Schurr, P. H., and Ingraham, F. D.: Myelomalacia and multiple cavitations of spinal cord secondary to adhesive arachnoiditis; experimental study. *A.M.A. Arch. Path.* 57: 138-146, 1954.
24. Gardner, W. J.: Anatomic anomalies common to myelomeningocele of infancy and syringomyelia of adulthood suggest a common origin. *Cleveland Clin. Quart.* 26: 118-133, 1959.
25. Morgagni, G. B.: *De Sedibus et Causis Morborum* 1761. Trans. by B. Alexander. London: A. Millar and T. Cadell, 1769.
26. von Recklinghausen, F.: Untersuchungen über die Spina bifida. *Arch. f. path. Anat.* 105: 243; 373, 1886.
27. Penfield, W., and Coburn, D. F.: Arnold-Chiari malformation and its operative treatment. *Arch. Neurol. & Psychiat.* 40: 328-336, 1938.
28. Feigin, I.: Arnold-Chiari malformation with associated analogous malformation of midbrain. *Neurology* 6: 22-31, 1956.