

Revised Classification of Posterior Fossa Cysts and Cystlike Malformations Based on the Results of Multiplanar MR Imaging

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MR and clinical data on 31 patients with posterior fossa CSF collections were analyzed. A clear separation of these patients into classical categories was not possible because of new information obtained from the MR images. We present a new classification of these disorders. The Dandy-Walker malformation, Dandy-Walker variant, and mega-cisterna magna seem to represent a continuum of developmental anomalies of the posterior fossa. A possible embryologic basis for this continuum is suggested. Discrete posterior fossa CSF collections that are clearly separate from the fourth ventricle and vallecula are classified as posterior fossa cysts. Posterior fossa CSF collections that communicate with the fourth ventricle and are associated with cerebellar atrophy are classified as prominent cisterna magna. Both the Dandy-Walker complex and posterior fossa cysts can cause enlargement of the posterior fossa and scalloping of the inner table of the occipital bone. The Dandy-Walker complex presents with seizures, developmental delay, and enlarging head size; it requires CSF diversion when associated with hydrocephalus. Posterior fossa cysts present with symptoms of a posterior fossa mass; they generally require surgical resection. Prominent cisterna magna is a result of degenerative disorders and requires no surgical therapy. This new classification facilitates both diagnosis and therapy of these disorders.

MR revealed that disorders previously referred to as the Dandy-Walker malformation, the Dandy-Walker variant, and the mega-cisterna magna actually are not separate entities, but appear to represent steps on a continuum of developmental anomalies of the posterior fossa. Because of this, we suggest a new term, the Dandy-Walker complex, be used to describe this continuum.

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In 1914, Dandy and Blackfan [1] described a 13-year-old patient with severe hydrocephalus and associated cystic dilatation of the fourth ventricle. Autopsy of this patient revealed, in addition to the ventricular enlargement, hypoplasia of the vermis, wide separation of the cerebellar hemispheres, dilatation of the aqueduct, and absence of the foramina of Luschka and Magendie. In the years since this original description, many cases of the so-called "Dandy-Walker malformation" (so named by Benda [2]) have been described, but definitions of the entity have been modified to include findings encountered in a particular case or series of cases [3-22]. All definitions include three features: (1) cystic dilatation of the fourth ventricle, (2) dysgenesis of the cerebellar vermis, and (3) a high position of the tentorium.

Radiologic characterization has been based on axial CT and, before that, pneumoencephalography. Invariably, differentiation between the Dandy-Walker malformation and other posterior fossa fluid collections has been difficult. Terms such as mega-cisterna magna, Dandy-Walker variant, and posterior fossa arachnoid cyst have been applied. Differentiation of these entities is often difficult [9, 14, 16, 18]; moreover, in the absence of associated anomalies, this differentiation may not be of clinical significance [21, 22]. MR imaging has been shown to be of considerable value in the diagnosis of congenital anomalies of the brain and in the

elucidation of their embryogenesis [23, 24]. However, we found that differentiation of the various groups of posterior fossa cysts was more difficult by MR than by CT. The wide variation in the appearance of the cerebellar vermis on sagittal images was particularly troublesome; varying degrees of hypoplasia, compression, and rotation of the vermis seemed to span all the classic categories. We reexamined posterior fossa CSF collections identified with multiplanar high-quality MR in the hope of better defining their classification and origin.

Materials and Methods

The MR scans of 31 patients with posterior fossa CSF collections were evaluated retrospectively. The 19 male and 12 female patients ranged in age from 6 days to 69 years (mean age, 13.5 years; median age, 11 years). Patients were referred for enlarging head size/hydrocephalus (13 patients), developmental delay (five), ataxia (six), seizures (four), dementia (two), and facial dysmorphism (one). Patients were included in the series if a CSF collection that was subjectively believed by the authors to be larger than normal was present in the posterior fossa.

Images were obtained with a GE 1.5-T system. Imaging parameters included a 256×256 acquisition matrix, 20-cm field of view, 5-mm section thickness, and interslice gap of 1–2.5 mm. Sagittal spin-echo (SE), 400–600/20/2 (TR/TE/excitations), sequences were obtained in 29 patients (two patients were not imaged in the sagittal plane). Axial SE, 2500–3000/30, 70, images were obtained in all patients. Axial or coronal SE, 600/20, images were obtained in 10 patients to better visualize certain aspects of the underlying disease or, in patients less than 6 months old, to assess brain maturation [25]. All axial images were obtained in a plane parallel to the canthomeatal line.

The cerebellar vermis and hemispheres were assessed for hypoplasia, atrophy, compression, rotation, and (when present) axis of rotation. The vermis was designated as hypoplastic or agenetic if the inferior vermian lobules were either small or absent, respectively (the normal vermis has nine lobules). Compression of the vermis was

diagnosed if frank deformity was observed or if the sulci between the folia were judged subjectively to be diminished in size. Rotation was detected as rotation of a line (from the most superior aspect of the culmen to the most inferior aspect of the uvula) out of approximate parallelism with the brainstem and into a more horizontal plane. Invariably, this rotation occurred by anterior displacement of the culmen and posterior displacement of the uvula (Fig. 1). Diminished size of the cerebellar hemisphere without enlargement of the sulci or fissures between the folia reflected hypoplasia. If all the lobules were present but the fissures between the lobules and folia were enlarged, the cerebellar hemisphere was considered to be atrophic rather than hypoplastic. The straight sinus normally makes an angle of approximately $50\text{--}75^\circ$ with respect to the superior sagittal sinus (personal observations). This "tentorial angle" was measured, and angles greater than 80° were one of several indicators of posterior fossa enlargement. Elevation of the tentorium and expansion of the occiput were also noted. The presence or absence, location, and size of the falx cerebelli were noted. The brainstem was assessed for size, displacement, and morphology. The degrees of ventricular dilatation and aqueductal patency were assessed. The inner table of the skull in the posterior fossa was examined for evidence of scalloping by the posterior fossa cyst. Finally, the supratentorial structures were examined for callosal anomalies, gyral anomalies, or any other disease.

Results

The patients were categorized into three main groups on the basis of clinical data and MR findings (Table 1): Dandy-Walker complex, prominent cisterna magna, and posterior fossa cysts. The Dandy-Walker complex was further divided into two subgroups, based on the presence or absence of the vermis at the mid-fourth ventricle on axial images.

The Dandy-Walker complex group comprised 19 patients who had posterior fossa CSF collections that showed clear-cut communication with the fourth ventricle (via the valleculla and fourth ventricular foramina) and no atrophy of the cere-

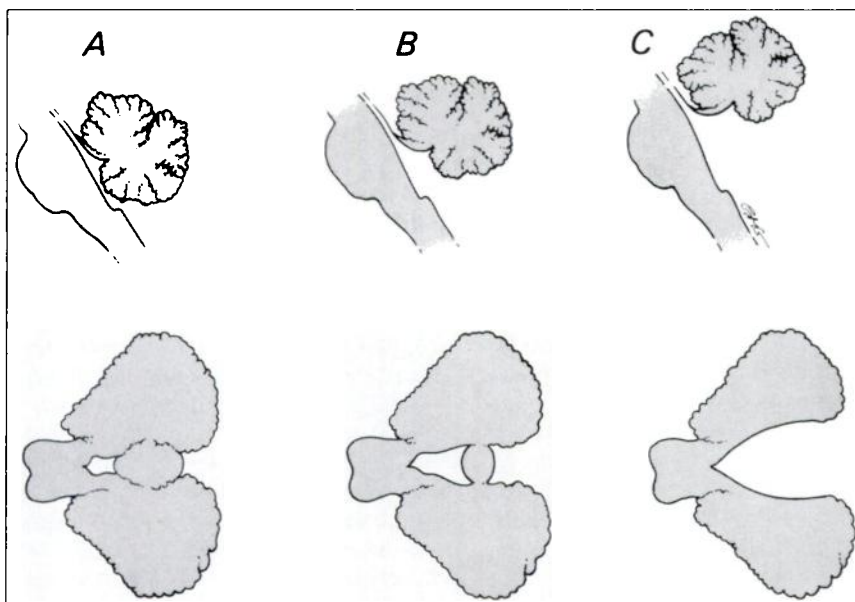


Fig. 1.—Schematic drawings show effect rotation of vermis has on appearance of fourth ventricle in axial plane.

A, Without rotation of vermis, axial images show vermian tissue interposed between fourth ventricle and cisterna magna.

B, With small amount of vermian rotation, axial cuts through level of fourth ventricle suggest some enlargement of fourth ventricle, but there is still vermian tissue interposed between ventricle and cisterna magna.

C, With large degree of vermian rotation, axial images through level of fourth ventricle suggest vermian hypoplasia or aplasia, despite presence of fully formed vermis.

TABLE 1: Dandy-Walker Groups Based on Clinical Data and MR Findings

Finding	No. of Patients			
	Dandy-Walker Complex		Prominent Cisterna Magna (n = 6)	Posterior Fossa Cysts (n = 6) ^c
	Vermis Absent (n = 12) ^a	Vermis Present (n = 7) ^b		
Enlarged posterior fossa	8	6	0	0
Hydrocephalus or CSF shunt	11	2	0	1
Developmental delay	3	3	2	0
Vermian atrophy	0	0	6	0
Vermian hypoplasia	10	4	0	0
Vermian compression	10	7	0	3
Vermian rotation	5	2	0	0
Supratentorial anomalies	8	1	0	0
Falx cerebelli present	5	7	5	6
Falx cerebelli midline	2	0	5	6
Abnormal straight sinus angle	6	3	0	0
Atrophy of cerebellar hemisphere	0	0	6	0
Hypoplasia of cerebellar hemisphere	12	2	0	0

^a "Classic" Dandy-Walker group (vermis absent on axial images).

^b Mega-cisterna magna group (vermis seen on axial images).

^c Posterior fossa arachnoid cyst group.

bellar hemispheres or vermis. Two subgroups were discernible: (1) Dandy-Walker type A, in which the vermis was absent on axial images at the level of the fourth ventricle because of rotation and/or hypoplasia or aplasia of the vermis (as observed on sagittal images); and (2) Dandy-Walker type B, in which the vermis was present at the level of the fourth ventricle on axial images and complete or nearly complete on sagittal images, and there was clear continuity of CSF collections with the fourth ventricle.

The prominent cisterna magna group comprised patients who had posterior fossa CSF collections with demonstrable communication with the fourth ventricle and cerebellar/vermian atrophy.

The group with posterior fossa cysts comprised patients with posterior fossa CSF collections without demonstrable communication with the fourth ventricle and no atrophy of the cerebellum.

Dandy-Walker Complex

Dandy-Walker complex type A.—All 12 patients in this subgroup had an enlarged fourth ventricle and the cisterna magna was not separated from the mid-fourth ventricle by an intervening vermis on axial images; standard definitions would classify this as a Dandy-Walker malformation. The six male and six female patients ranged in age from 1 week to 17 years (average age, 4.6 years) at the time of the MR studies. All patients had macrocephaly; three exhibited developmental delay and one had seizures.

The cerebellar hemispheres were hypoplastic and the vermis was hypoplastic or absent in all patients (Fig. 2). The hemispheres ranged in size from 8 mm in the largest dimension (in an 11-year-old) to 4 × 4 × 4 cm. By inspection, the hemispheres had a normal ratio of gray to white matter and, where it could be evaluated, a grossly normal lobular pattern.

The vermis was completely absent in two patients. In the remaining 10 patients the vermian dysplasia ranged from mild hypoplasia of the inferior lobules to a barely detectable remaining superior vermis. In five patients, the vermis was rotated (Figs. 3 and 4) and in 10 patients the vermis appeared compressed. In all the patients in whom the vermis was only slightly hypoplastic, it was severely rotated, permitting free communication between the fourth ventricle and cisterna magna, and had a "classic" Dandy-Walker appearance on axial images (Fig. 4). In two patients with partial vermian hypoplasia, the inferior portions of the cerebellar hemispheres were closely apposed beneath the hypoplastic vermis, creating the appearance of a normal fourth ventricle and normal vermis (Fig. 5).

In eight patients, the tentorium and torcular herophili were high and the posterior fossa was enlarged. The inner table of the occipital bone was scalloped. In six of these patients, the normal 50–75° acute angle formed by the junction of the straight sinus with the superior sagittal sinus was instead obtuse, ranging from 90° to 150°. In the other two patients the angle between the straight and superior sagittal sinuses was normal; the tentorium was elevated anteriorly rather than posteriorly and the torcular was normal in position. One of the four patients without elevation of the tentorium had had a shunt for many years and exhibited marked thickening of the occipital bone, suggesting that the posterior fossa had once been enlarged but had diminished relative to the remainder of the brain (Fig. 6). Scalloping of the calvaria was not seen in the four patients with normal posterior fossa size.

The falx cerebelli, most easily seen on the long TR/long TE sequence, was identified in five patients. It was midline in two patients, displaced to the side of the small hemisphere in three patients, and elongated (3 cm long) in two of the latter patients.

Eleven of the 12 patients had hydrocephalus. Shunt cath-

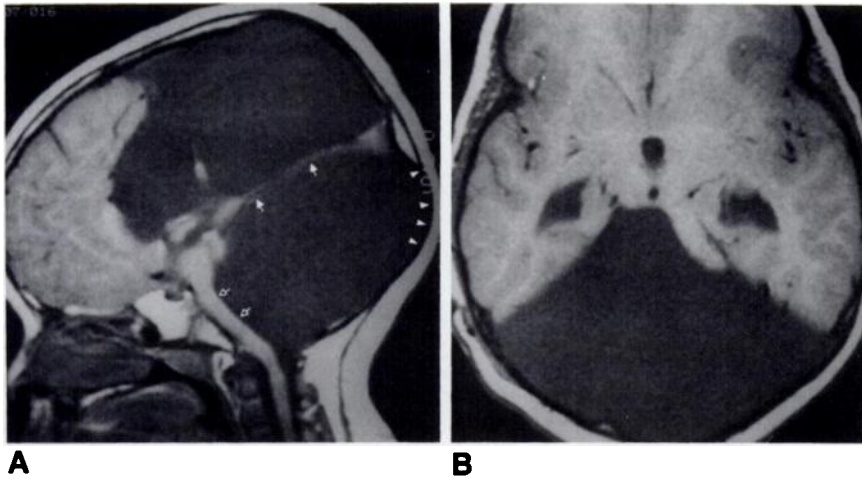


Fig. 2.—Classic Dandy-Walker deformity type A with associated dysgenesis of corpus callosum and interhemispheric cyst.

A, Midline sagittal spin-echo MR image, 600/20, shows tentorium elevated (*solid arrows*), brainstem compressed against clivus (*open arrows*), and inner table of occipital bone scalloped (*arrowheads*). Cerebellar vermis is absent.

B, Axial spin-echo MR image, 600/20, reveals dilatation of temporal horns of lateral ventricles. Left cerebellar hemisphere is extremely hypoplastic. Right cerebellar hemisphere, not seen on this image, was seen to be quite hypoplastic on lower images.

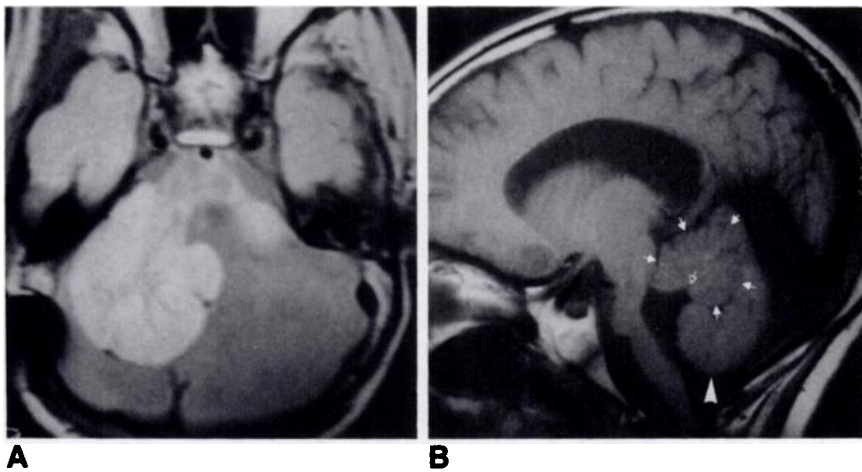


Fig. 3.—Dandy-Walker deformity (type A) with rotation of cerebellar vermis.

A, Axial spin-echo MR image, 2500/30, at level of mid-fourth ventricle shows severe hypoplasia of left cerebellar hemisphere and absence of vermis at this level.

B, Midline sagittal spin-echo MR image, 600/20, shows intact vermis (*solid arrows*) that is rotated approximately 90° counterclockwise around axis at level of inferior aqueduct. Note location of fastigium of vermis (*open arrow*). Posterior fossa is only slightly enlarged (straight sinus is slightly high) and straight sinus angle (see text) is normal. Note normal position of right cerebellar tonsil (*arrowhead*).

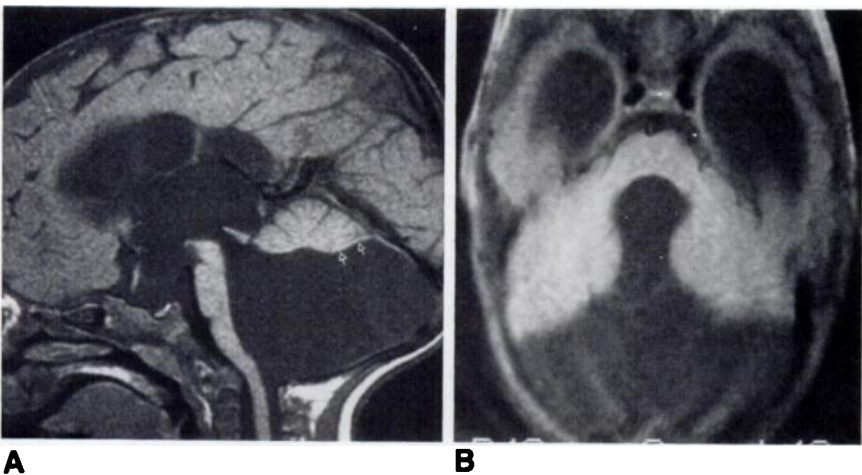


Fig. 4.—Dandy-Walker deformity (type A) with marked dilatation of aqueduct.

A, Midline sagittal spin-echo MR image, 600/20. Vermis is slightly hypoplastic (six lobules) but markedly rotated superiorly. Note intact inferior medullary velum (*arrows*). This patient has an enlarged posterior fossa, compressed brainstem, dilated aqueduct, and agenesis of corpus callosum.

B, Axial spin-echo MR image, 600/20, at level of mid-fourth ventricle shows continuity of fourth ventricle and cisterna magna. On axial images at this level or lower this would be misdiagnosed as vermian aplasia.

eters were present in the posterior fossa cyst in six patients; three had shunts in the lateral ventricles as well. The cerebral aqueduct could not be visualized in the six patients with shunts. Of the five patients without CSF diversion, the aqueduct was enlarged in one (with severe hydrocephalus), normal in two (with mild hydrocephalus), and stenotic in two (severe

hydrocephalus). In two of the patients with shunts and one with severe hydrocephalus, the aqueduct appeared compressed by the anteriorly and superiorly displaced vermis (Fig. 7).

The brainstem was normal in four patients. In six patients, it was flattened as a result of compression against the clivus

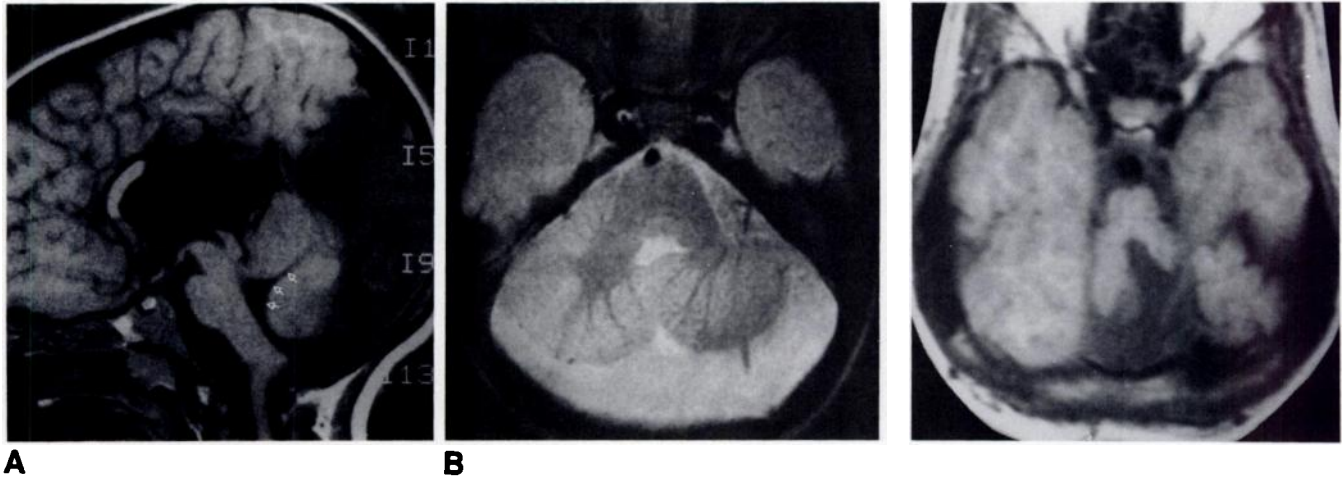
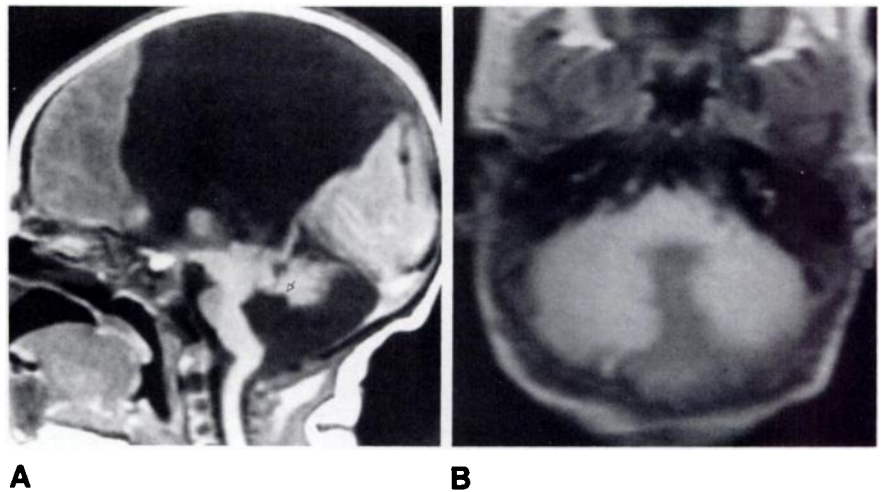


Fig. 5.—Dandy-Walker deformity (type A) with associated dysgenesis of corpus callosum.
A, Midline sagittal spin-echo MR image, 600/20, reveals enlarged posterior fossa CSF collection and probable elevated tentorium. Vermis is hypoplastic; however, appearance of apparently normal vermian body and fourth ventricle is present because of apposition of inferior cerebellar hemispheres in midline below vermian body (*arrows*).
B, Axial spin-echo MR image, 2800/70, at level of inferior fourth ventricle shows apposition of cerebellar hemispheres in midline without intervening vermian body, verifying interpretation of anatomy shown in **A**.

Fig. 6.—Dandy-Walker deformity (type A) after many years of diversion of CSF from posterior fossa cyst. Axial spin-echo MR image, 600/20, reveals marked thickening of occipital bone and small posterior fossa. Note extreme hypoplasia of cerebellar hemispheres. Vermian body was not seen on any axial images through the fourth ventricle.

Fig. 7.—Dandy-Walker deformity (type A) with abnormal flexure at craniocervical junction.
A, Midline sagittal spin-echo MR image, 600/20, shows compression, hypoplasia, and counter-clockwise rotation of vermian body (note location of fastigium [*arrow*]). Rotated vermian body seems to compress mesencephalic tectum, possibly contributing to aqueductal stenosis. Cervicomedullary junction forms an angle of 110° .
B, Axial spin-echo MR image, 600/20, at mid-fourth ventricle shows continuity of ventricle with cisterna magna at this level.



(Fig. 2A). In two patients there was an abnormal flexure at the craniocervical junction, forming an angle of approximately 110° between the medulla and the upper cervical spinal cord in one patient (Fig. 7) and an angle of approximately 120° in the other. Normally this angle ranges from approximately 135° to 180° [26].

Associated anomalies were present in eight patients. Six patients had dysgenesis of the corpus callosum (complete agenesis in two and the presence of the genu only in three and the genu and anterior body in one). Two of the patients with callosal anomalies had a large interhemispheric cyst; a third had subependymal gray-matter heterotopias. The seventh patient had a large hemangioma in the soft tissues of the neck; the eighth had a cardiac ventricular septal defect. One of the patients with complete agenesis of the corpus

callosum and the patient with the hemangioma had a normal-sized posterior fossa.

Dandy-Walker complex type B.—The seven patients in this subgroup were separated from the type A patients by interposition of a portion of the cerebellar vermian body between the mid-fourth ventricle and the enlarged cisterna magna on axial images. This malformation traditionally would be considered the mega-cisterna magna. This subgroup comprised four males and three females 2 weeks to 23 years old (mean, 9 years). Two patients had developmental delay, one had developmental delay and seizures, two had signs of hydrocephalus, one had dysmorphic facial features, and one had a unilateral sixth-nerve palsy.

The inferior lobules of the cerebellar vermian body were hypoplastic in four patients (Fig. 8); the vermian body was complete in the

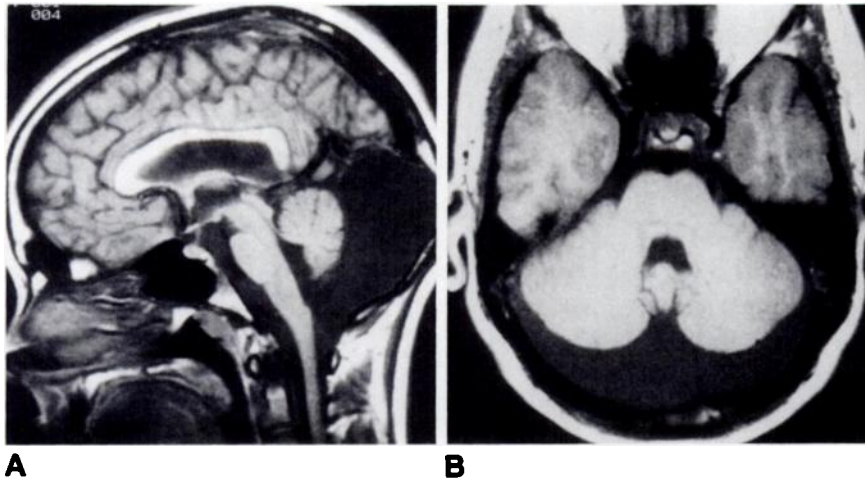


Fig. 8.—Dandy-Walker deformity (type B).

A, Midline sagittal spin-echo MR image, 600/20, reveals hypoplasia of inferior vermis, dilatation of inferior fourth ventricle, and enlarged posterior fossa. Vermis is rotated approximately 15–20° in a counterclockwise direction. Scalloping of occipital bone and abnormal straight sinus angle are present. In classic terms, these findings would be categorized as a mega-cisterna magna, but note the similarities to Dandy-Walker deformity (type A). This patient was severely developmentally delayed.

B, Axial spin-echo MR image, 600/20, through fourth ventricle shows inferior vermis separating ventricle from cisterna magna. (Compare with Fig. 1.) Such a deformity would have seemed clearly separable from Dandy-Walker deformities (type A) on axial CT or pneumoencephalography.

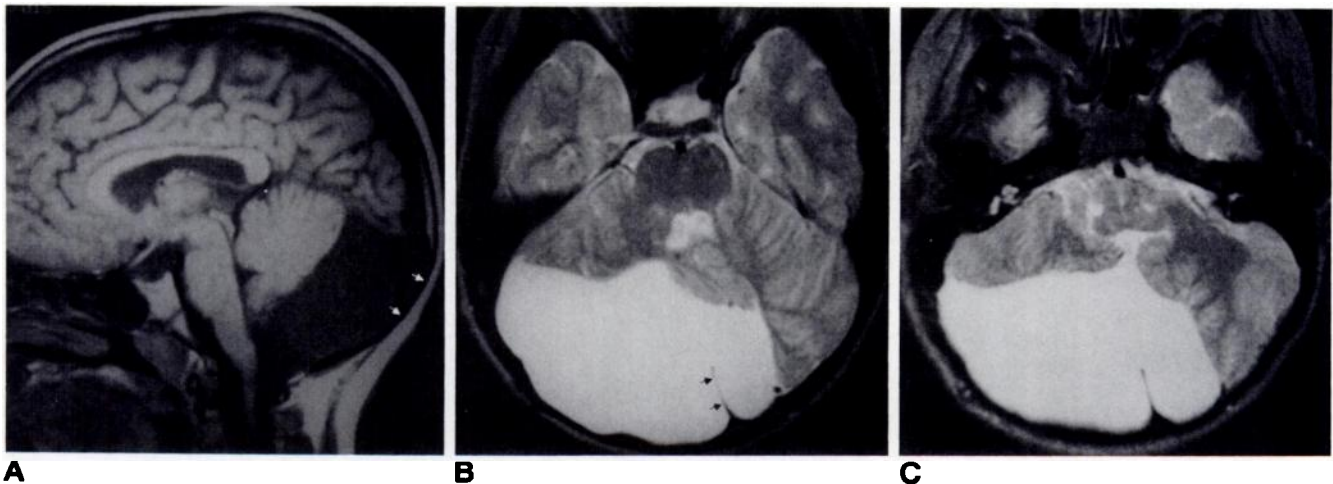


Fig. 9.—Dandy-Walker deformity (type B).

A, Midline sagittal spin-echo MR image, 600/20, shows compression of inferior vermis by retrovermian CSF collection. Inner table of occipital bone is scalloped (arrows).

B, Axial spin-echo MR image, 2500/70, at level of mid-fourth ventricle. Inferior vermis and ventricle are displaced. Right cerebellar hemisphere is smaller than left. Falx cerebelli is elongated and displaced to right (arrows).

C, Axial spin-echo MR image, 2500/70, at level of vallecule suggests continuity of posterior fossa CSF collection with fourth ventricular outflow foramina through vallecule.

other three. The vermis was compressed in all seven patients. Slight rotation of the vermis was evident in two patients. In one patient the inferior vermis was displaced anteriorly; this anomaly would classically be classified as an arachnoid cyst, but images clearly showed communication with the fourth ventricle via the vallecule (Fig. 9). The cerebellar hemispheres were mildly hypoplastic in two patients and normal in five. The inferior fourth ventricle was enlarged in all four patients with vermian hypoplasia.

The posterior fossa was enlarged to some degree with scalloping of the inner table of the occipital bone in all seven patients. An upward bulging of the cisterna magna through the tentorial incisura, elevating the posterior tentorium but maintaining a normal straight sinus angle, was seen in three patients. The straight sinus angle was reversed in a fourth patient, coursing upward from the vein of Galen to the torcular and forming an obtuse angle with the superior sagittal sinus. The angle of the straight sinus was approximately 90° in two

patients with a normal sagittal sinus and tentorium anteriorly, but an upward bowing of the sinus posteriorly by superior herniation of the cisterna magna.

The falx cerebelli was present but displaced from the midline in all seven patients. The length was less than 1.5 cm in all patients.

Hydrocephalus was present in three patients; a CSF diversion catheter had been placed from the lateral ventricles in two patients. One patient had metopic suture synostosis and a dysgenic corpus callosum (hypoplastic splenium). The aqueduct was of normal size and the brainstem was normal in all patients in this group. There were no other associated anomalies.

Posterior Fossa Cysts

This group comprised six patients with discrete posterior fossa CSF collections that did not communicate directly with

the fourth ventricle. The three males and three females in this group were 12–69 years old (mean, 38 years). Five patients had ataxia; one of these had headaches and another demonstrated dysdiadochokinesia as well. The sixth patient had chronic schizophrenia; the cyst was an incidental finding. Of the five patients with ataxia, four were found at surgery to have arachnoid cysts; the fifth improved after cyst-peritoneal shunting, and the ataxia was presumed to have been caused by an arachnoid cyst. The cyst in the schizophrenic patient was not treated.

The vermis and hemispheres were complete and of normal size in all patients. Displacement of the vermis and ipsilateral hemisphere anteriorly by the cyst was observed in four patients (Figs. 10 and 11). The cysts were midline in two patients and to the right of midline in the other four.

The brainstem was normal in three patients and slightly displaced and compressed in three patients. The size of the posterior fossa, the position of the tentorium and straight sinus, the falx cerebelli, the ventricular size, the cerebral aqueduct, and the supratentorial structures were normal in all patients. There were no associated systemic anomalies. A focal scalloping of the inner table of the occipital bone (adjacent to the cyst) was seen in three of the patients.

Prominent Cisterna Magna

All six patients in this group had an enlarged cisterna magna without enlargement of the posterior fossa. The patients in this group were all males, ranging in age from 3 to 47 years (average, 21.2 years). Three patients presented with dementia, two with developmental delay, and one with ataxia. No patients in this group had hydrocephalus or CSF-diverting shunts.

Both the vermis and the cerebellar hemispheres were obviously atrophic with marked prominence of the sulci and fissures (Fig. 12). There was no evidence of vermian hypoplasia, rotation, or compression.

The falx cerebelli was seen in five of the six patients; it was small (less than 1.5 cm) and in the midline. The brainstem was atrophic in one patient and normal in five. Associated anomalies included supratentorial atrophy in three patients and supratentorial infarcts in two. The posterior fossa size,

straight sinus angle, ventricular size, and cerebral aqueduct were normal in all patients in this group.

Discussion

Embryologic Development of the Cerebellum

At about the fifth week of gestation, a thickening occurs bilaterally in the alar plate of the rhombencephalon, forming the rhombic lips, which are the primordia of the cerebellar hemispheres. The glial and neuronal cells that compose the cerebellum migrate to their final locations in the cerebellar hemispheres by two general pathways. The neurons of the deep cerebellar nuclei and the Purkinje layer of the cerebellar cortex migrate radially outward from the germinal matrix in the wall of the fourth ventricle (Fig. 13A) [27–29]. Generation of these neurons in the germinal matrix seems to occur between 9 and 13 weeks of gestation [27]. The neurons of the granular layer of the cerebellar cortex, in contrast, have a more complicated journey. At 11–13 weeks, these cells begin to migrate tangentially from a germinal zone (in the lateral portion of the rhombic lips) over the cerebellar surface, forming the transitory external granular layer [27]. The cells in this external layer then proliferate at a high rate as the cerebellum begins a rapid growth period (commencing in the 13th gestational week and continuing until the seventh postnatal month). The proliferation of the external granule cells continues as, at 16 weeks, daughter cells begin to migrate inward (Fig. 13B). Some of the daughter cells form the basket and stellate cells of the outer (molecular) layer of the cerebellar cortex. Others continue their migration and move inward past the Purkinje cells to form the inner granular layer of the cortex. The external granular layer attains maximum cell number in the first few postnatal months, then diminishes in size as the granule cells migrate inward. By the end of the first postnatal year, the external granular layer has essentially disappeared and the cerebellar cortex achieves its adult three-layered histologic composition—the outer molecular layer, middle Purkinje layer, and inner granular layer.

The afferent and efferent connections of the cerebellar cortex consist of fibers that synapse with the deep cerebellar nuclei and fibers that communicate with the rest of the central

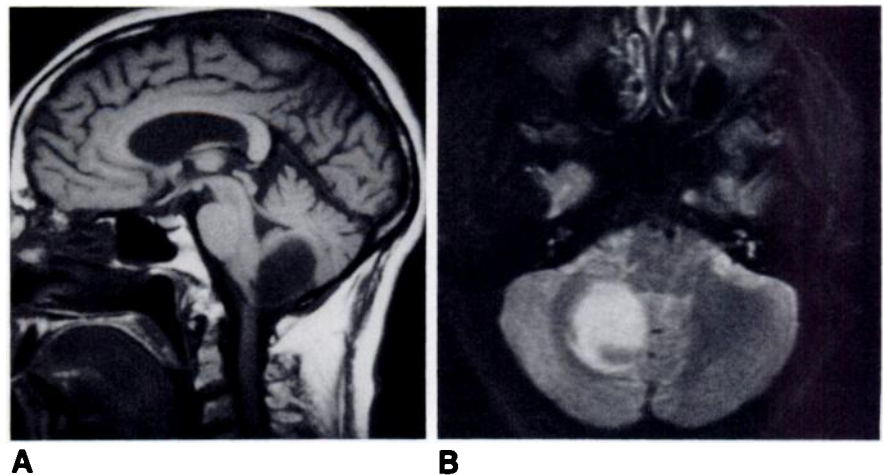


Fig. 10.—Retrocerebellar/intracerebellar posterior fossa arachnoid cyst (surgically proved).

A, Midline sagittal spin-echo MR image, 600/20, shows discrete CSF collection impressing on vermis and fourth ventricle.

B, Axial spin-echo MR image, 2500/70, shows mild brainstem compression as well. This patient had progressive ataxia.

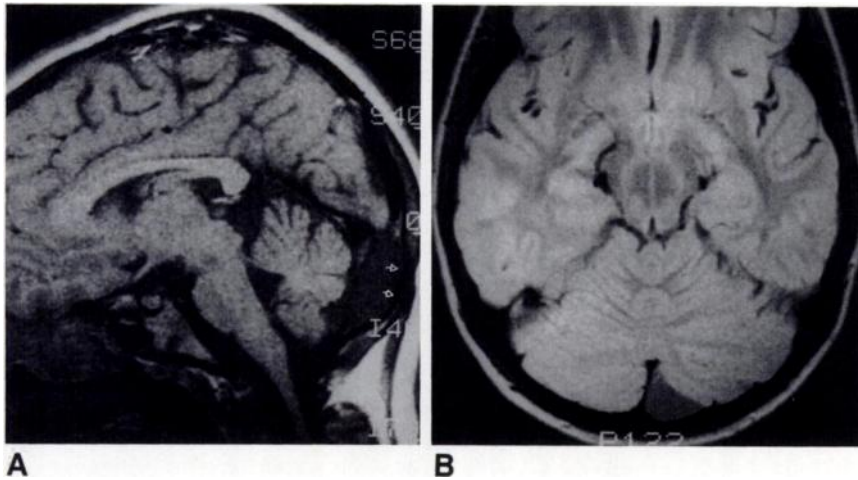


Fig. 11.—Posterior fossa cyst.
A, Midline sagittal spin-echo MR image, 600/20, shows small retrovermian CSF collection. Scalloping of adjacent inner table of occipital bone is present (*arrows*). CSF collection is clearly separate from vallecula and fourth ventricle.
B, Axial spin-echo MR image, 2500/30, reveals minor mass effect on left cerebellar hemisphere by presumed arachnoid cyst.

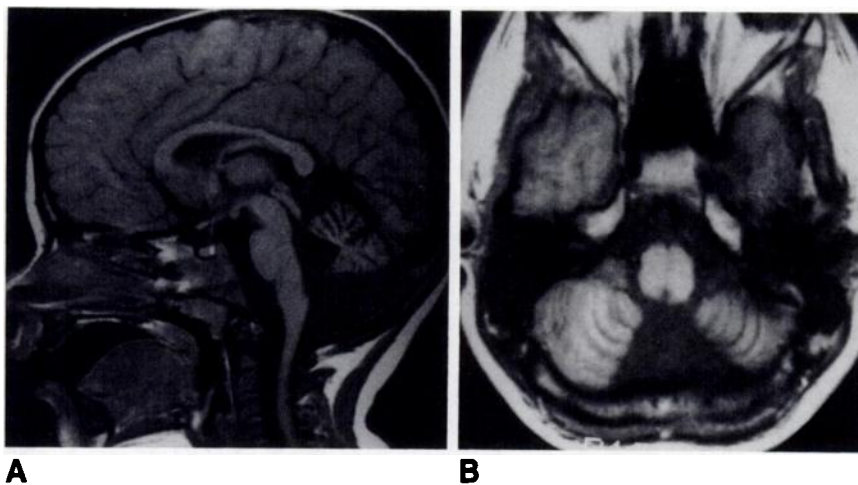


Fig. 12.—Prominent cisterna magna.
A, Midline sagittal spin-echo MR image, 600/20, shows vermian atrophy (diminished size of folia with resulting sulcal prominence) and consequent enlargement of cisterna magna and fourth ventricle.
B, Axial spin-echo MR image, 600/20, reveals sulcal prominence of cerebellar hemisphere resulting from diffuse cerebellar atrophy. Cisterna magna and perimedullary cisterns are quite prominent.

nervous system via the superior, middle, and inferior cerebellar peduncles. Cortical afferent and efferent white-matter tracts seem to form even if the migration of some of the cerebellar cortical components is interrupted [27, 30]. As a result of the formation of these white-matter tracts, the cerebellum is grossly normal in appearance (with a cortex and medulla that are roughly proportionate to those in a normal cerebellum), despite the developmental insult. The cerebellum appears merely hypoplastic no matter what type of insult occurs [30].

The cerebellar vermis forms from the fusion of the developing hemispheres. The fusion begins when the hemispheres meet superiorly in the midline during the ninth gestational week, and it continues inferiorly as the hemispheres grow, with the entire vermis being formed by the end of the 15th week [31].

One other important concept in the development of the cerebellum as it relates to posterior fossa cysts is the complex development of the roof of the fourth ventricle. Bonnevie and Brodal [32] have shown that on the 11th fetal day in the normal mouse, the roof of the fourth ventricle is divided by a ridge of developing choroid plexus into anterior and posterior membranous areas (Fig. 14). By the end of the 11th day, the

anterior membranous area (above the choroid ridge) is incorporated into the developing choroid plexus. The posterior membranous area (below the ridge of choroid) remains; an area within it eventually cavitates to form the midline foramen of Magendie. The lateral foramina of Luschka open at some later and as yet unknown time.

The most intriguing aspect of our findings was the difficulty in clearly differentiating with MR the Dandy-Walker malformation from what is usually called a mega-cisterna magna. These traditionally have been treated as clearly distinct clinical and radiographic entities. We placed all patients with clear continuity of the fourth ventricle and enlarged cisterna magna on axial images in the Dandy-Walker type A subgroup, and we placed those patients with an intervening inferior vermis in the Dandy-Walker type B subgroup. The latter subgroup is usually referred to as the mega-cisterna magna. This distinction could be accomplished on axial images; however, when the sagittal images were reviewed, it was difficult to make the separation. One reason for the difficulty was the fact that two of the patients in the Dandy-Walker type A subgroup (Fig. 3) had a mild degree of vermian hypoplasia and cisterna magna enlargement that was quite similar to that in two patients in the Dandy-Walker type B subgroup (Fig. 8). The

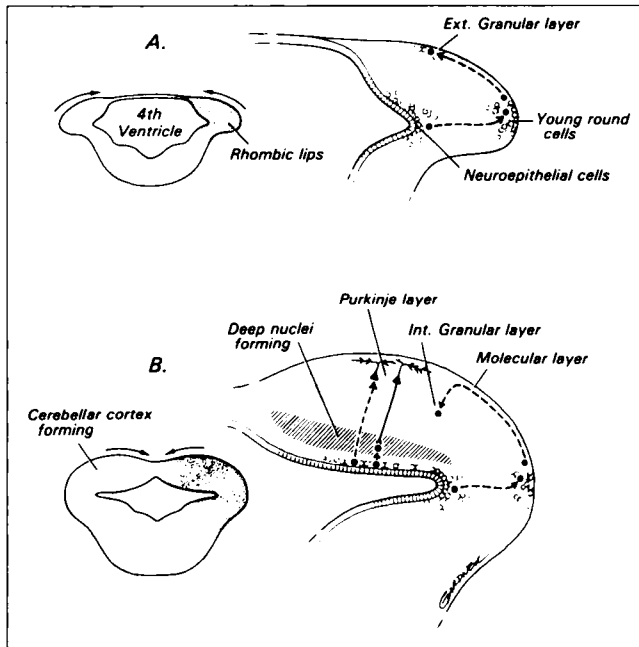


Fig. 13.—Schematic drawing shows normal formation of cerebellum.

A, During fifth week of gestation, thickening occurs bilaterally in ear plate of rhombencephalon, forming rhombic lips that are primordia of cerebellar hemispheres. At 11–13 weeks, cells that will form granular layer of cerebellar hemisphere begin to migrate tangentially from germinal zone in lateral portion of rhombic lips over cerebellar surface, forming transitory external (Ext.) granular layer.

B, At approximately 16 weeks of gestation, cells of external granular layer begin to migrate inward to form internal (Int.) granular layer of cerebellar cortex. These cells will eventually synapse with cells within deep cerebellar nuclei. Cells that will form Purkinje layer of cerebellar cortex and deep cerebellar nuclei originate in wall of fourth ventricle. These cells migrate directly from germinal matrix to their final destinations within cerebellar hemispheres. Cortical afferent and efferent white-matter tracts seem to form even if migration of some cerebellar cortical components is interrupted. As a result of the formation of these white-matter tracts, cerebellum is grossly normal in appearance (but hypoplastic), even if developmental insult occurs.

major difference was that a marked upward rotation of the vermis created an appearance of continuity in the type A patients. On axial images, we were unable to differentiate vermian rotation from vermian hypoplasia (Fig. 1). Enlargement of the posterior fossa also overlapped in the two subgroups.

The Dandy-Walker patients in the type B subgroup all had posterior fossa enlargement; however, in the type A patients there was a wide range of posterior fossa sizes and tentorial elevation. In several patients, the appearance of a large posterior fossa was more a function of the hypoplasia of the cerebellar hemispheres and vermis than actual enlargement.

It was also impossible to separate these patients on clinical grounds. Nine (75%) of the Dandy-Walker patients in the type A subgroup had an enlarging head caused by hydrocephalus and have developed normally, six after CSF shunting and three without intervention. Two Dandy-Walker patients (29%) in the type B subgroup had hydrocephalus; both have done well after CSF diversion. This high percentage of good results substantiates the encouraging prognosis for patients with

Dandy-Walker malformations in other recent reports [21, 22]. In each of the two Dandy-Walker subgroups, three patients had nonspecific developmental delay and one had seizures. The more severe clinical conditions correlated with the presence of supratentorial anomalies (gray-matter heterotopias, dysgenesis of the corpus callosum with interhemispheric cyst). This correlation has been reported in other patients also [21, 22]. The severity of the clinical symptoms did not appear to correlate with the severity of the hindbrain deformity.

Although hypoplasia of the vermis is frequently mentioned, the severity of the hypoplasia of the cerebellar hemispheres has not been emphasized in the literature; this hemispheric hypoplasia was striking in many of our cases (Figs. 1 and 5). At least one of the hemispheres was severely hypoplastic in half of the Dandy-Walker patients in the type A subgroup; the cerebellar hemispheres of all patients in this group were at least slightly hypoplastic. Some hypoplasia of the hemispheres was obvious in two of the seven Dandy-Walker patients in the type B subgroup.

Cerebellar hypoplasia and/or a fourth ventricle–cisterna magna cyst were present in all patients in both Dandy-Walker subgroups. However, there was no apparent relationship between the severity of the two findings: some patients had marked cerebellar hypoplasia with a small cyst, some had mild hypoplasia with a large cyst, while still others had marked hypoplasia and a large cyst. Therefore, it is reasonable to suggest that two separate processes were at work in the development of this group of malformations.

From the earlier discussion of the embryogenesis of the cerebellum, it is apparent that hypoplasia of the cerebellar hemispheres as observed by MR is a nonspecific result of interference with the normal migration of either the Purkinje cells from the germinal matrix in the roof of the fourth ventricle or the young neurons (that eventually form the granular layer of the cerebellar cortex) from the rhombic lips. Since formation of the hemispheres begins before the ninth gestational week and continues until the seventh postnatal month, an insult that might cause cerebellar hypoplasia is difficult to localize temporally. It seems reasonable to assume that severe hypoplasia may occur as the result of an early or a severe insult; if so, mild hypoplasia would result from a late or a mild insult.

Formation of the posterior fossa cyst is the other process that is occurring in the development of these malformations; this event may be timed more easily. Some background information may be helpful in understanding this process. The fourth ventricle outlet foramina of Magendie (the medial aperture) and Luschka (the lateral apertures) are inconsistently patent in patients with the Dandy-Walker malformation. In general, the lateral apertures are more commonly patent than the medial aperture [17]. Hirsch et al. [17] have proposed that the cause of the posterior fossa cyst is delayed or nonopening of the median aperture with consequent enlargement of the fourth ventricle, which then bulges posteriorly to form the fourth ventricle–cisterna magna complex. They suggest that this bulging fourth ventricular complex hinders formation of the inferior vermis. The longer the opening of the median aperture is delayed, the larger the posterior fossa

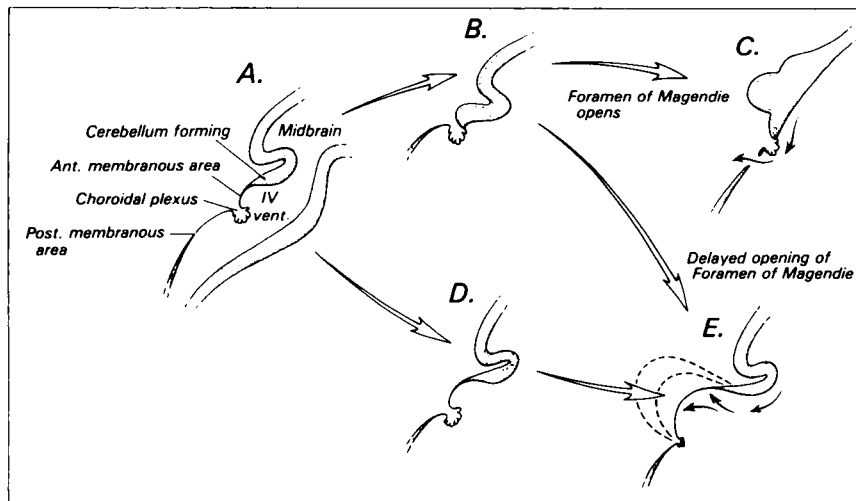


Fig. 14.—Normal and abnormal development of the fourth (IV) ventricle.

A, Early in gestation, roof of fourth ventricle is divided by ridge of developing choroidal plexus into anterior (Ant.) and posterior (Post.) membranous areas.

B, Normally, anterior membranous area is incorporated into developing choroid plexus.

C, Posterior membranous area remains and eventually cavitates to form midline foramen of Magendie.

D and E, If anterior membranous area is not incorporated into developing choroid plexus (D) or if there is a delayed opening of the foramen of Magendie, the roof of the fourth ventricle can balloon posteriorly to form a fourth ventricular-cisterna magna cyst (E).

cyst becomes, and, according to this theory, larger cysts cause greater enlargement of the posterior fossa and more prominent hypoplasia of the cerebellar vermis. Hart et al. [10] have clearly shown, however, that in some patients both the median and lateral apertures are patent. Moreover, the presence of certain systemic anomalies in some patients (such as cardiac septal defects and polydactyly) necessitates the occurrence of an insult prior to late in the eighth to early in the 10th gestational week, when the foramen of Magendie normally opens [31]. Bonnevie and Brodal [32] (cited in [18]) and Kalter [33] have shown that in some mutant mice the anterior membranous area in the roof of the fourth ventricle (see discussion of embryologic development) does not incorporate into the developing choroid plexus; instead, it persists and bulges posteriorly as a dilatation of the fourth ventricle, stretching and attenuating the inferior vermis. The final result is a defect identical to the Dandy-Walker malformation [32, 33].

We believe that these patients suffer an insult to both the developing cerebellar hemispheres and the developing fourth ventricle. In some patients, the insult is severe and diffuse and results in a large fourth ventricle cyst (which causes an enlarged posterior fossa) and a severely hypoplastic cerebellum. This is the classic Dandy-Walker malformation. In other patients, the insult is more localized. If the developing cerebellum is predominantly involved, the result can be cerebellar hypoplasia (hemispheric and/or vermian) without marked expansion of the posterior fossa, a condition that has been labeled the Dandy-Walker variant [12]. If the fourth ventricle is more involved, the result is an expanded posterior fossa with a large CSF collection that communicates with the fourth ventricle, but relatively minor hypoplasia of the cerebellum. This is the classic mega-cisterna magna. The difference in the degree of enlargement of the fourth ventricle (and the posterior fossa) presumably relates to the severity of the insult to the developing anterior membranous area or to the extent of delay of the opening of the foramen of Magendie; a more severe insult or a more delayed foraminal opening will result in a larger posterior fossa cyst and a larger posterior fossa. The cause of these events could be chromosomal

injury if the loci for the fourth ventricle roof and the cerebellum are in proximity. Alternatively, the insults could be to the developing brain itself.

The difficulty that we encountered in differentiating the classic Dandy-Walker malformation patients from those in the mega-cisterna magna group was not surprising because the differentiation of the various posterior fossa cystic malformations has always been difficult, both clinically and radiographically. With the use of pneumoencephalography, differentiation between the Dandy-Walker malformation and the mega-cisterna magna depended on visualizing the cerebellar vermis between the fourth ventricle and the cisterna magna [14, 16]. We have shown in this study that this apparent distinction may be merely the result of the degree of upward rotation of the vermis (Fig. 13). Although we cannot offer proof, it seems reasonable to conclude that the degree of vermian rotation may reflect the etiology of the fourth ventricle-cisterna magna complex; that is, the vermis may rotate when the fourth ventricle abnormality results from a lack of regression of the anterior membranous area but not when delayed opening of the median aperture is the cause or vice versa. Further work may help elucidate these mechanisms.

Differentiation of a Dandy-Walker malformation from a retrocerebellar posterior fossa arachnoid cyst by pneumoencephalography was said to be dependent on the speed with which air moved into the CSF collection from the fourth ventricle. Lack of filling or slow filling was believed to be diagnostic of an arachnoid cyst. Such a differentiation was shown to be dependent on technique [14, 16]. The patient shown in Figure 9 would have been classified as having a posterior fossa arachnoid cyst by classical criteria because of the separation of the fourth ventricle from the cisterna magna on axial images and the displacement of the vermis and fourth ventricle by the CSF collection. However, both axial and sagittal images indicate clear communication between the fourth ventricle, vallecule, and cisterna magna in this patient, whereas a discrete cyst membrane or separation from the vallecule is seen in the patients classified as having arachnoid cysts (Figs. 10 and 11). We cannot conclusively prove noninvasively that a small velum is not present between

the vallecule and the retrocerebellar CSF collection and that the patient shown in Figure 9 does not, in fact, have a retrocerebellar arachnoid cyst as opposed to a mega-cisterna magna. However, whether or not free communication exists between the retrocerebellar CSF and the fourth ventricle, the treatment of this patient (or any similar patient) will not be affected. The patient, if symptomatic because of hydrocephalus or compression of the cerebellum, will be treated by a cyst-peritoneal CSF diversion. In this way, retrocerebellar arachnoid cysts differ from intracerebellar arachnoid cysts (such as that illustrated in Fig. 10), which often are treated by surgical extirpation. Moreover, we see displacement of the vermis, similar to that seen in Figure 9, by the CSF collection in other disorders that are clearly part of the Dandy-Walker complex. Because the treatment, prognosis, and clinical and radiologic appearances are so similar to the other anomalies in the group, it is reasonable to include an anomaly such as the one illustrated in Figure 9 within the Dandy-Walker complex.

The clinical and radiographic evidence in this study indicates that the Dandy-Walker malformation, the Dandy-Walker variant, and the mega-cisterna magna are part of an overlapping spectrum that have a related embryologic origin and a similar clinical prognosis. Moreover, all patients with this spectrum of disorders are treated conservatively unless they have hydrocephalus, in which case a CSF diversion catheter is placed. It is therefore proposed that these terms be abandoned and replaced by a single entity called the Dandy-Walker complex. Although we found a similar clinical spectrum for all patients within this category, a slightly higher incidence of associated anomalies (and hence a worse clinical prognosis) might be expected for the patients with both cerebellar and fourth ventricular anomalies, because the presence of both anomalies suggests a more severe or more diffuse insult to the fetus.

In two patients with vermian hypoplasia, the midline sagittal image showed an intact vermis and a normal fourth ventricle because of apposition of the inferior cerebellar hemispheres below the vermis. This "pseudo-fourth ventricle" has been described in the pneumoencephalographic literature [9, 14]; the observer who fails to note the absence of normal vermian folia inferiorly (Fig. 3) can be misled. The pseudo-fourth ventricle appearance is particularly prominent after shunting of the posterior fossa cyst.

One other interesting observation in this study is that the falx cerebelli is inconsistently present as well as elongated and displaced in patients with the Dandy-Walker complex. The falx cerebelli was present in only 12 of 19 patients and its length ranged up to 3 cm (Fig. 9). The normal falx cerebelli generally is less than 1 cm long. Moreover, the falx was always displaced from the midline, usually to the side of the smaller hypoplastic hemisphere. The embryologic events leading to this displacement and to the elongation of the falx cerebelli are not clear.

The patients in the prominent cisterna magna group were largely those with degenerative diseases of the CNS. The appearance of the atrophic cerebelli in this group was far different from that of the hypoplastic cerebelli in the Dandy-Walker complex. The cerebellar cortex was thin, the folia

small, and the sulci and fissures between the folia enlarged (Fig. 12). It was interesting that those patients with marked cerebellar hypoplasia had no symptoms referable to the posterior fossa, whereas those with cerebellar atrophy had cerebellar signs. Clearly, in the absence of a normally formed cerebellum, the developing brain forms compensatory pathways that perform the functions of the cerebellum, whereas the mature brain, on losing existing cerebellar tracts, compensates much more slowly, if at all.

In all patients in the arachnoid cyst group, the cyst was localized and clearly separate from the apertures of the fourth ventricle and the vallecule. Definite mass effect on the adjacent cerebellar hemisphere was seen in three of the four patients. We believe that the presenting symptoms of these patients (referable to the posterior fossa), the separation of the cystic collections from the fourth ventricle and vallecule, and the absence of posterior fossa atrophy can be used to clearly separate these patients from those with the other anomalies described in this communication. Moreover, the treatment (surgical resection of the cyst) differentiates these lesions from the Dandy-Walker complex and the prominent cisterna magna.

Our series did not include any patients with primary cerebellar agenesis, as described by Joubert et al. [34] and Boltshauser et al. [35]. Patients with primary cerebellar agenesis have nearly total aplasia of the cerebellar vermis; dysplasias and heterotopias of the cerebellar nuclei; nearly total absence of the pyramidal decussation; and anomalies in the structure of the inferior olivary nuclei, descending trigeminal tract, solitary fascicle, and dorsal column nuclei. Clinically, these patients are characterized by episodic hyperpnea, abnormal eye movements, ataxia, and mental retardation. The disorder is clearly genetic, because the reported cases have all occurred in a few families [35]. Although the MR appearance of primary cerebellar agenesis syndrome has not been described, the CT findings are characteristic. Absence of the vermis results in a pyramid-shaped fourth ventricle inferiorly and a bat wing appearance superiorly. The cerebellar hemispheres appose one another in the midline. The superior cerebellar peduncles are distinctly visualized as they extend superiorly toward the midbrain, surrounded by CSF. Large CSF collections are not present in the posterior fossa in these patients. In view of its genetic transmission, characteristic clinical presentation, and absence of a large CSF collection, primary cerebellar agenesis syndrome is clearly separate from the categories in our classification. Moreover, because the disorder is genetic, and therefore the result of a chromosomal anomaly, our theory of embryogenesis is not applicable.

Scalloping of the inner table of the calvaria is a classic finding in an arachnoid cyst and was seen in three of the six patients with arachnoid cysts in this study. Our finding of scalloping of the inner table of the occipital bone in 14 of 18 patients with the Dandy-Walker complex suggests that, in the posterior fossa, such scalloping is not useful as a differential factor.

We had hoped to identify associated abnormalities of the brainstem in those patients with cerebellar hypoplasia or atrophy. Specifically, hypoplasia of the olivary and red nuclei have been described in association with cerebellar hypoplasia

and the Dandy-Walker malformation [10, 11]. Unfortunately, the red nuclei are not well seen on MR scans of children, probably because of the small amount of iron in young brains; furthermore, the olivæ are extremely difficult to identify in 5-mm sections through the brainstem in young children. As a result, only gross anomalies, such as compression of the brainstem against the clivus by the large posterior fossa cyst, were identified.

In summary, posterior fossa CSF collections can result from abnormal development of the cerebellum or fourth ventricle, from degeneration of the cerebellum, or from formation of arachnoid cysts. We have presented a new classification of these disorders. The Dandy-Walker malformation, the Dandy-Walker variant, and the mega-cisterna magna seem to represent a continuum of developmental anomalies of the posterior fossa and cannot and need not be clearly separated clinically or anatomically. We suggest that these terms be abandoned and replaced by a single term, the Dandy-Walker complex. Some previously so-called posterior fossa arachnoid cysts are a part of this complex as well.

Discrete posterior fossa cystic collections are easily characterized because they are clearly separate from the fourth ventricle and cisterna magna on axial and sagittal images. Posterior fossa CSF collections that are continuous with the vallecula and cisterna magna and are associated with cerebellar atrophy are caused by the atrophy and are appropriately called prominent cisterna magna. The clinical presentation and symptoms of these three groups of cystic lesions are clearly different. This new classification facilitates both radiologic diagnosis and clinical treatment.

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