Radiat Med (2006) 24:471–481
DOI 10.1007/s11604-006-0049-7

MINIREVIEW

Midline cystic malformations of the brain: imaging diagnosis and classification based on embryologic analysis

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Accepted: February 7, 2006

Abstract This article describes a classification and imaging diagnosis of intracranial midline cystic malformations based on neuroembryologic analysis. Midline cystic malformations are classified into two categories from an embryologic point of view. In one category, the cyst represents expansion of the roof plate of the brain vesicle, and in the other the cyst consists of extraaxial structures such as an arachnoid membrane or migrating ependymal cells. Infratentorial cysts, such as the Dandy-Walker cyst or Blake's pouch cyst, and supratentorial cysts, such as a communicating interhemispheric cyst with callosal agenesis or a dorsal cyst with holoprosencephaly, are included in the first category. Infratentorial arachnoid cavities, such as the arachnoid cyst, arachnoid pouch, and mega cisterna magna, are in the second category. Noncommunicating interhemispheric cysts, such as interhemispheric arachnoid cyst or ependymal cyst, with callosal agenesis are also in the second category. A careful review of embryologic development is essential for understanding these midline cysts and for making a more accurate radiologic diagnosis.

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This work was presented as an educational lecture at the 64th Japan Radiological Society meeting (April 2005) in Yokohama.

Key words Dandy-Walker cyst · Blake's pouch cyst · Arachnoid pouch · Interhemispheric cyst with callosal agenesis · Dorsal cyst with holoprosencephaly

Introduction

Over the past century, various definitions and classification systems have been devised regarding midline cystic malformations of the brain.[1–4] Since the introduction of magnetic resonance imaging (MRI), these cystic malformations of the brain, especially in the posterior fossa, have been found with greater frequency than was previously suspected. Despite the superiority of MRI for identifying and characterizing these malformations, there is still a great deal of confusion regarding their classification, terminology, and spectrum of expression.[5–8] This confusion is best resolved by a more complete understanding of human neuroembryology.[9,10] Familiarity with the normal embryonic development of the brain is helpful for understanding the pathogenesis and imaging appearance and for categorizing various terminologies that have been used to describe midline cystic malformations of the brain.

To understand the pathogenesis of midline cystic malformations, we first review the normal embryologic development of the rhombencephalon and prosencephalon and then present their typical imaging features and illustrate the distinguishing clues for their differential diagnosis.

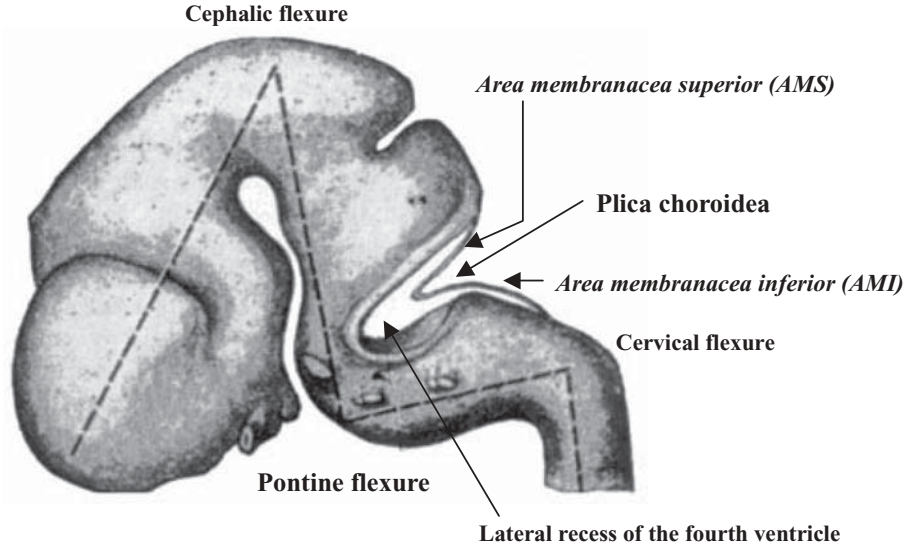


Fig. 1. Developing roof of the rhombencephalon. The thin roof of the rhombencephalon becomes folded and forms a transverse crease (plica choroidea), which divides the membranous roof of the fourth ventricle into superior and inferior membranous areas (area membranacea superior and inferior). The choroid plexus of the fourth ventricle will develop in the plica choroidea by apposition of the ependymal mesoderm

Posterior fossa cysts

Development of the rhombencephalon and surrounding mesenchymal tissue

As early as Carnegie stage 9 [approximately 20 days postovulation (p.o.)], the neural tube divides into three primary brain vesicles: the prosencephalon, mesencephalon, and rhombencephalon. The prosencephalon develops into the telencephalon (future cerebral hemispheres and striatum) and diencephalon (future thalamus and hypothalamus), and the rhombencephalon develops into the metencephalon (future pons and cerebellum) and myelencephalon (medulla oblongata). During stage 14, the neural tube becomes bent in three regions: at the mesencephalon (cephalic flexure), at the cervicomedullary junction (cervical flexure), and in the metencephalon (pontine flexure).[11–13] After curvature of the pons (pontine flexure), the roof plate of the rhombencephalon becomes wider and diamond-shaped. It is formed of ependymal cells that are reinforced on the outer surface by pia mater resulting from differentiation of the mesenchymal tissue of the meninx primitiva.[14] This differentiation is accompanied by vascular development and by the formation of a transverse vascular fold that divides the roof plate of the rhombencephalon into a rostral part (area membranacea superior) and a caudal part (area membranacea inferior).[15] The transverse vascular fold, which is also called the plica choroidea, invaginates into the lumen of the rombencephalon (future fourth ventricle) and constitutes a rudimentary choroid plexus that extends from one lateral recess to the other (Fig. 1).[12] The cerebellum also begins to develop during stage 14 as two symmetrical thickenings on either side of the midline of the rhombencephalon.[16] These cerebellar bulges, called rhombic lips, arise in the dorsal alar plate just lateral to the rhombencephalic isthmus, where cells contribute to form the cerebellar vermis.[17] Thus, the cerebellar hemispheres unite at the midline by cells derived from the isthmus to form the beginning of the vermis. With continued development by intense cell multiplication in the cerebellar cortex, the area membranacea superior disappears with the formation of the cerebellar vermis (superior and inferior medullary vellum), and the choroid plexus becomes attached to the caudal edge of the cerebellum.[9,10] On the other hand, the area membranacea inferior, which consists of an ependymal membrane that is histologically different from the cellular lining of the area membranacea superior, initially expands and forms a small diverticulum (called Blake's pouch), which eventually disappears, leaving a median aperture that becomes the foramen of Magendie (Fig. 2).[14,18] Blake[19] and Wilson[20] observed that dehiscence of the ependymal membrane becomes apparent at 7–8 weeks p.o. The foramen of Luschuka, which is found at the distal end of the lateral recess, appears later: Although the exact timing of opening is unknown, it is probably between 14 and 17 weeks p.o.[18]

At the same time that these changes occur in the rhombencephalic roof plate, the meninx primitiva, which surrounds the neural tube, differentiates. The loose mesenchymal tissue continues to thin out by progressive expansion of the extracellular space and gradual differentiation of the juxtaneural (future pia mater) and peripheral (future dura mater) cellular layers. This rarefaction of cells in the meninx primitiva is an intrinsic process inherent in the development of the meningeal tissue and occurs without any intervention from the

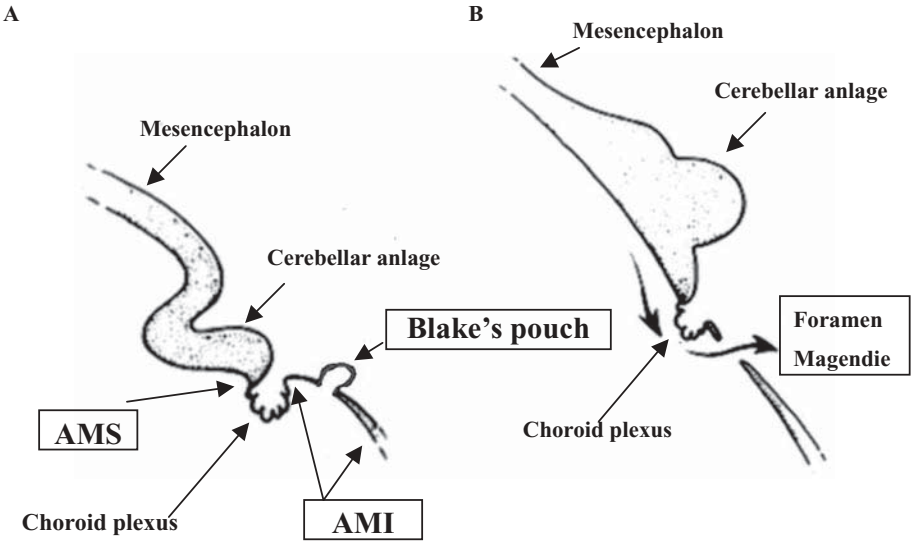


Fig. 2A,B. Developing choroid plexus and roof of the fourth ventricle. **A** Craniocaudal formation of the vermis leads to disappearance of the area membranacea superior (AMS), and the choroid plexus comes to lie at the caudal edge of the cerebellum. The area membranacea inferior (AMI) evaginates to create Blake's pouch. **B** The apex of Blake's pouch is normally thin and disappears, creating the foramen Magendie of the fourth ventricle

intraventricular fluid. Although Weed[15] believed that the intraventricular fluid under excessive pressure broke through the tela choroidea of the fourth ventricle, thereafter gradually dissecting the meningeal space and then forming the subarachnoid space, it is now thought to be the liquid of the pericerebral space that develops independent of the ventricular system.[21] Therefore, the intraventricular and extraventricular fluids may coexist separately until they are connected by apertures in the fourth ventricle (at 7–8 weeks p.o.).[14] The cisterna magna, which is the portion of the wide subarachnoid space below the inferior cerebellar surface, appears within a few days after the foramen Magendie opens. Continuity between the intra- and extraventricular liquid compartments is established by apertures that open in the tela choroidea of the fourth ventricle that are derived from the area membranacea inferior. The development of the subarachnoid cisterna magna by hollowing out of the meninx primitiva and the formation of the foramen of Magendie by attenuation of the area membranacea inferior are active processes, intrinsic to each structure. Thus, contrary to Weed's opinion, the intraventricular and extraventricular fluids are independent of any mechanical action of the ventricular fluid.

When the cisterna magna appears, the lateral portions of the dura mater leaflets, which later become the tentorium cerebelli, are visible.[14] These lateral parts of the tentorium merge on the midline in a craniocaudal sequence and later form the superior sagittal sinus, the straight sinus, the torcular, and the transverse sinus: The tentorium is apparent by the end of the embryonic period, and the torcular herophili is in its normal position by approximately 12 weeks p.o.[18]

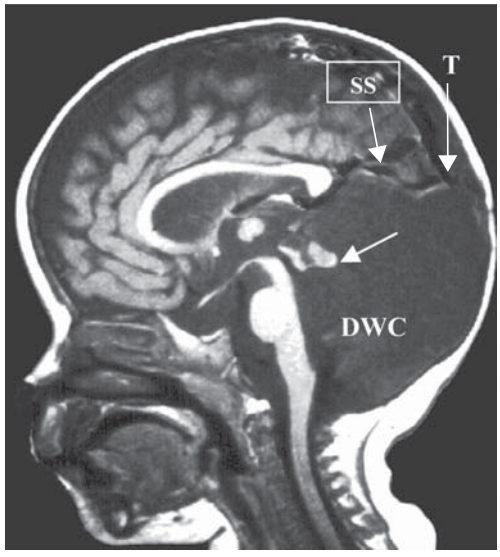


Fig. 3. Dandy-Walker cyst from a 1-year-old female infant. Midsagittal T1-weighted image shows a hypoplastic vermis with a markedly enlarged primitive fourth ventricle, a so-called Dandy-Walker cyst (*DWC*). A high-positioned torcular (*T*) with an elevated straight sinus (*SS*) is also noted

Cysts representing expansion of the rhombencephalic roof plate

Dandy-Walker cyst

The Dandy-Walker cyst (DWC) (Fig. 3), which characterizes Dandy-Walker malformation,[3] represents expansion of the primitive fourth ventricular roof due to persistence of the area membranacea superior (AMS).

Because persistence of the AMS may result from incomplete formation of the cerebellar vermis, the Dandy-Walker malformation always presents an absent or markedly hypoplastic vermis.[22] Although the imaging manifestations of this malformation can vary, a typical case has a huge DWC with a large posterior fossa and elevated transverse and straight sinuses and torcular herophili, which may result from the developmental arrest of the tentorium cerebelli secondary to existence of the distended DWC.[10] No fourth ventricular choroid plexus is identifiable in a DWC. Therefore, identification of a normal choroid plexus in the inferior medullary vellum is important to rule out a Dandy-Walker malformation.[18] On the other end of the spectrum is a mildly hypoplastic vermis with a large fourth ventricle filling a normal-sized posterior fossa with a torcular herophili in a normal position: These features are usually called a Dandy-Walker variant. The term Dandy-Walker variant was first introduced by Harwood-Nash and Fitz[23] to describe posterior evagination of the tela choroidea of the fourth ventricle associated with a mildly hypoplastic vermis. Raybaud[14] used this term to describe a malformation with varying degrees of hypoplastic vermis with expansion (often considerable) of the fourth ventricle, which communicates freely with the subarachnoid space, and reserved the term Dandy-Walker malformation for cases in which there is no communication between the dilated fourth ventricle and the subarachnoid space. However, it now looks as though this posterior evagination of the tela choroidea may indicate a distended Blake's pouch (Blake pouch cyst), as mentioned below, and it should be excluded from the spectrum of Dandy-Walker malformation.[18]

Blake's pouch cyst

Blake's pouch refers to transient outpouching of the area membranacea inferior (AMI), which becomes the tela choroidea of the fourth ventricle, before the foramen of Magendie opens (Fig. 2A).[24–26] Blake's pouch cyst is defined as persistent cystic evagination of the AMI.[18,27] Therefore, Blake's pouch cyst is located in a widened cerebellar vallecula underneath the inferior vermis (Fig. 4). In contrast to the Dandy-Walker malformation, the cerebellar vermis is formed normally but may be displaced superiorly; it often looks like a hypoplastic vermis. The cyst does not extend beyond the internal occipital protruberance, so the torcular herophili is usually in a normal position. Blake's pouch cyst is occasionally accompanied by internal hydrocephalus (dilatation of the whole ventricular system), probably due to an incomplete aperture of the foramen of Magendie.[14] In addition, in some cases, the fourth ventricular choroid plexus may be identified as it elongates along the superior cyst wall under the inferior surface of the vermis.[18,28]

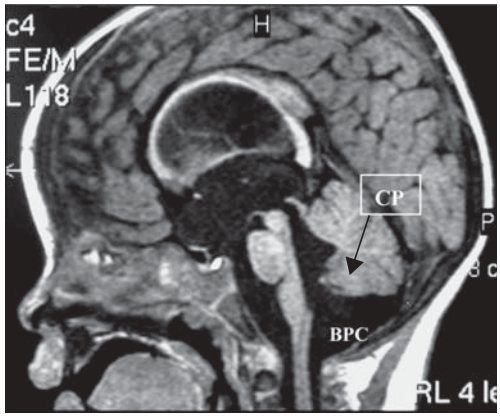


Fig. 4. Blake's pouch cyst in a 1-year-old male infant. Midsagittal T1-weighted image shows a small cystic cavity indicating Blake's pouch cyst (*BPC*) under the normally formed inferior vermis. The choroid plexus (*CP*) of the fourth ventricle is identified as it elongates along the inferior surface of the vermis. Dilatation of the whole ventricular system, which may indicate noncommunicating hydrocephalus, is also shown

Extraaxial cysts consisting of arachnoid membrane

Arachnoid cyst

Arachnoid cysts are developmental variants of the meninx primitiva that surrounds the neural tube during differentiation of the mesenchyma. They are cerebrospinal fluid (CSF) collections that develop within the layers of the arachnoid membrane. Therefore, a posterior fossa arachnoid cyst develops in the posterior aspect of the rhombencephalic roof plate and does not communicate freely with the fourth ventricle or perimedullary subarachnoid space. Because the cerebellar vermis is formed normally but is usually displaced anteriorly by the cyst, the fourth ventricular choroid plexus and inferior medullary velum are also displaced anteriorly (Fig. 5). In addition, these cysts are occasionally accompanied by noncommunicating supratentorial hydrocephalus due to compression of the fourth ventricle. If a huge cyst develops during the early fetal period, the tentorium cerebelli may be dehisced posteriorly, and the torcular herophili is then formed in a high position, as in the Dandy-Walker malformation.[27]

Arachnoid pouch

An arachnoid pouch is the same as an arachnoid cyst pathologically. We use the term "arachnoid pouch" when the retrocerebellar arachnoid cyst communicates freely with the subarachnoid space and does not displace

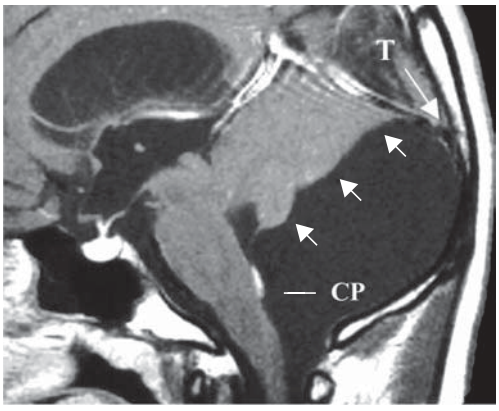


Fig. 5. Arachnoid cyst in a 16-year-old girl. Midsagittal T1-weighted postcontrast image shows a cyst located inferior to a normally formed but inferiorly compressed vermis (*arrows*). Note the enhanced choroid plexus (*CP*) compressing the anterior wall of the cyst. A high-positioned torcular (*T*) is also shown

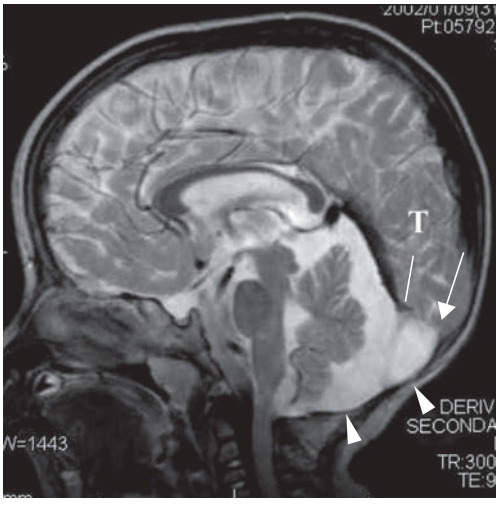


Fig. 6. Arachnoid pouch in a 3-year-old male child. Midsagittal T2-weighted image shows a retrocerebellar cyst bulging above the internal occipital protruberance (*arrows*) toward the superior cerebellar cistern to cause remodeling of the inner table of the occipital bone (*arrowheads*). No compression of the cerebellum or fourth ventricle is noted. Dysgenetic torcular (*T*) is also shown

the cerebellum anteriorly.[14] The posterior fossa is usually large, and the pouch extends to the superior cerebellar subarachnoid space beyond the internal occipital protruberance (Fig. 6). Dehiscence in the posterior tentorium cerebelli with a high-positioned torcular is occasionally seen.[27]

Mega cisterna magna

The term "mega cisterna magna" was first used by Gonsette et al.[29] to describe a grossly enlarged retrocerebellar subarachnoid space that they ascribed to

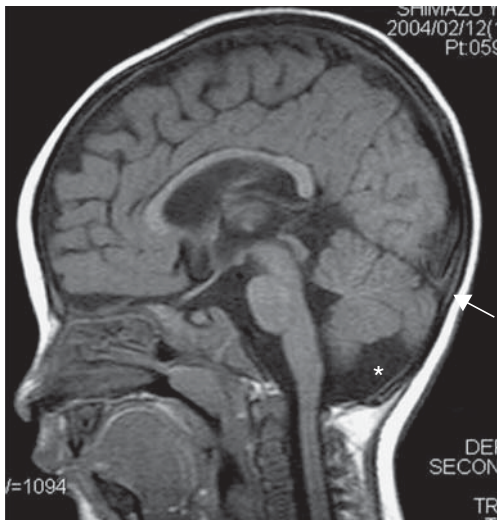


Fig. 7. Mega cisterna magna in a 3-month-old male infant. Midsagittal T1-weighted image shows an enlarged subarachnoid space below the inferior cerebellar surface (*asterisk*), which exceeds the anatomical limits of the cisterna magna but does not extend beyond the internal occipital protruberance (*arrow*)

cerebellar atrophy in a series of adult patients. Since then, this term has been loosely applied to a large retrocerebellar CSF-containing space with a normal vermis and cerebellar hemisphere, such as in a arachnoid pouch mentioned above. However, morphologically, the mega cisterna magna is a developmental variation of the posterior fossa characterized by expansion of the cisterna magna, in which the height usually extends 2.5 cm up from the foramen magnum along the inner table of the occipital bone to a point midway between the posterior lip of the foramen magnum and the internal occipital protruberance; the depth is usually 5 mm and the width about 2 cm (Fig. 7).[30,31] However, it should also be noted that in the neonatal population the cisterna magna is usually considered to be comparatively larger than that in adults, as the cerebellum continues to grow for several months after birth.[10,14] Thus, because there are no definitive measurements for the size of the cisterna magna, a diagnosis of mega cisterna magna cannot be based solely on its size. To distinguish mega cisterna magna from arachnoid pouch, it is important to know the anatomical limits of the cisterna magna, which must not extend beyond the internal occipital protruberance.[27]

Supratentorial cysts

Development of the prosencephalon and surrounding mesenchymal tissue

The prosencephalon can be horizontally subdivided into the rostral part of the telencephalon and the caudal part

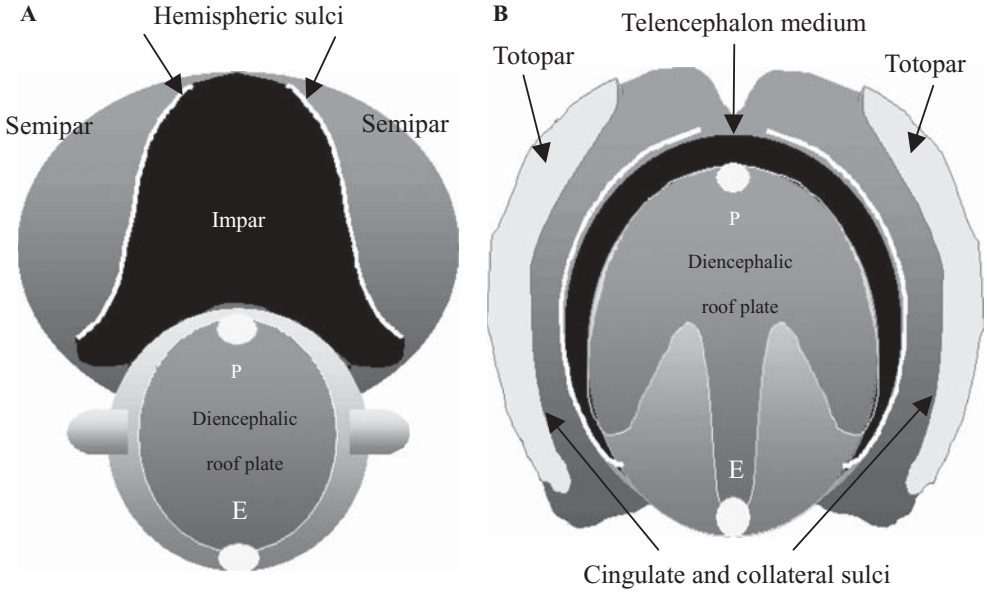


Fig. 8A,B. Hemispheric cleavage. **A** Dorsal view of the holospheric telencephalon (three-vesicular stage). Two hemispheric sulci (*white lines*) appear in the dorsal wall of the telencephalic holosphere. They divide the dorsal wall of the holosphere into three zones: one median zone (telencephalon impar) between two sulci and bilaterally two paramedian zones (telencephalon semipar). *P*, paraphysis; *E*, epiphysis (pineal body). **B** Dorsal view of the hemispheric telencephalon (five-vesicular stage). The hemispheric outpocketings begin to develop in the paramedian zones. The cingulate and collateral sulci on the medial walls of the growing hemisphere demarcate the telencephalon semipar from the completely paired telencephalon totopar. *P*, paraphysis; *E*, epiphysis (pineal body)

of the diencephalon.[12] By stage 13–14 (approximately 33 days p.o.), the telencephalon is a smooth median vesicle and represents a prospective hemisphere; but at this stage it is a nonpaired holosphere that consists of a primordial telencephalon "impar." Normally, in stage 15 (approximately 35 days p.o.), two hemispheric sulci (future hippocampal sulci) appear in the dorsal wall of the telencephalic holosphere near the paraphysis (a small evagination located in the most rostral part of the diencephalic roof plate) and converge rostrally and ventrally toward the lamina terminalis. Therefore, these sulci divide the dorsal wall of the holospheric telencephalon into three zones: one median zone (telencephalon "impar") between the two sulci telencephalou "impar" and bilaterally two paramedian zones (telencephalon "semipar"), which are paired dorsally but are continuous ventrally through the lamina reuniens (Fig. 8A). After that, completely paired zones, which consist of anlages of the neocortices (telencephalon "totopar"), develop in the paramedian zones. The telencephalon "totopar" undergoes differential cell proliferation to produce two expanding "hemispheres" (Fig. 8B). Owing to rapid development of the telencephalon "totopar," the telencephalon "impar" and "semipar" are displaced toward the limbus (margin) of the hemispheric vesicle, eventually forming the limbic system.[32,33] Moreover, because of their rapid growth, which exceeds that of the

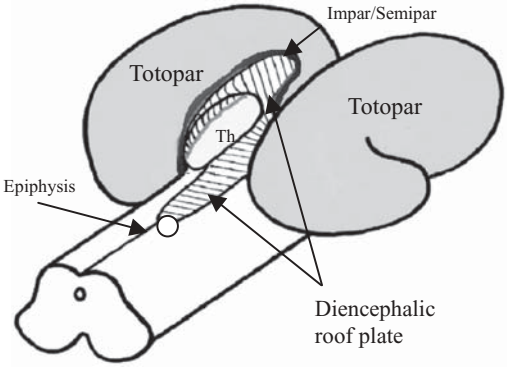


Fig. 9. Folding of the diencephalic roof plate. Due to rapid development of the telencephalon totopar, the telencephalon impar and semipar are displaced toward the limbus (margin) of the hemispheric vesicle. Moreover, the diencephalic roof plate is folded by expansion of the hemispheric vesicles and covers the thalamic eminences (*Th*)

diencephalon, the diencephalic roof plate is folded by expansion of the hemispheric vesicles and covers the thalamic eminences bilaterally (Fig. 9).[34]

At stage 15, the initial portion of the interhemispheric fissure develops over the most rostral and midline telencephalon of the "lamina reuniens" of His (also called the telencephalon medium) (Fig. 8B), where the telencephalic commissural fibers, such as the corpus callosum,

hippocampal commissure, and anterior commissure, will develop.[35,36] Thus, this area (lamina reuniens) is the anlage of all the "allocortical" formation of the rhinencephalon and also promotes the ingrowth of developing commissural fibers. Therefore, we hereafter refer to it as the commissural plate. During stages 15–23 (35–57 days p.o.), the interhemispheric fissure develops further posteriorly and deepens as a result of disproportionate growth of the evaginating cerebral hemispheres and midline telencephalic structures, such as the lamina terminalis and commissural plate.[21,37]

At the same time that the interhemispheric fissure is developing posteriorly, the commissural plate is growing posteriorly, and the interhemispheric fissure is becoming filled by mesenchyma, which forms the permissive substrate (callosal precursor) for axonal crossing of the commissural plate and later differentiates into the falx cerebri, anterior cerebral arteries, and subarachnoid space.[35,36] The choroid plexus of the lateral ventricle is formed by epithelial lamina, which is derived from the velum transversum at the transition to the diencephalic roof plate (epithelial roof of the third ventricle), together with meninx from the interhemispheric fissure.[38]

At the end of the embryonic period (stage 23), pioneer fibers of the corpus callosum pass through the dorsal part of the lamina reuniens (Fig. 10), where the massa commissuralis is formed as the callosal precursor. Eventually, the callosal fibers, which emanate from the isocortical plate of the hemispheric vesicles, begin to decussate into the massa commissuralis—in the genu at approximately 11 weeks and then in the body, splenium and rostrum at 18–20 weeks of gestation.[35,36] Thus, callosal formation is naturally closely related to neocortical development of the telencephalon "totopar." The gross shape of the corpus callosum is apparent by the end of the fifth month of gestation. Thereafter, it grows continuously until maturity. The crescentic shape of the corpus callosum, as it appears in a midsagittal section, as well as its relation to the structures adjacent to it, may be readily explained by crescentic expansion of the hemispheres. The inferior margins of the corpus callosum, which become stretched around the thalami, cover the diencephalic roof plate (roof of the third ventricle). Moreover, with growth of the callosal body, the hippocampal formation is cleaved ventrally and dorsally, pushing the fornices under it and taking the gray indusium on its back. The hippocampal formation stretched by the developing corpus callosum seems to become the septum pellucidum. The hippocampal commissure is also carried backward by the developing corpus callosum and forms the psalterium (fornicial commissure) of the adult brain.[35]

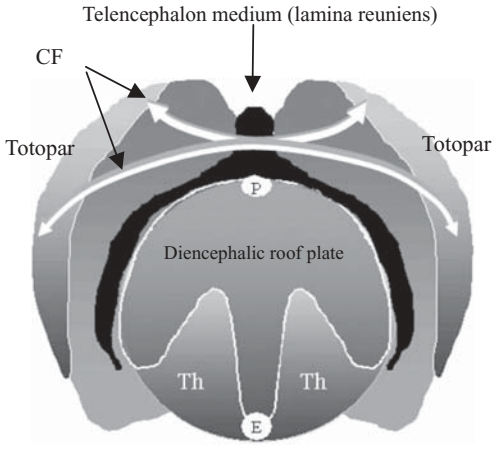


Fig. 10. Developing corpus callosum. The callosal fibers (*CF*) emanate from the developing telencephalon totopar and pass through the telencephalon medium (lamina reuniens), which is located in the median and most rostral part of the hemispheric telencephalon. *P*, paraphysis; *E*, epiphysis (pineal body); *Th*, thalamic eminence

Cysts representing expansion of the diencephalic roof plate

Communicating interhemispheric cyst with callosal agenesis

In 1973, Probst,[39] after an extensive evaluation of pneumoencephalography, reported superior extension of the roof of the third ventricle into the interhemispheric fissure in 40 of 50 children with partial or complete callosal agenesis; he called this anomaly communicating interhemispheric cyst (Fig. 11). He also divided these communicating interhemispheric cysts into "primary cysts," defined as being associated with a deficient falx cerebri, and "secondary cysts," in which the falx was intact. The developmental mechanism of "primary cysts" is still unclear. One possible explanation is that because the rudimentary corpus callosum during early gestation is located only in the anterior aspect of the third ventricle (and does not cover the roof of the third ventricle) fetal hydrocephalus can easily elevate the roof of the third ventricle toward the vertex and may impair formation of the falx, which is derived from the primitive meninx in the interhemispheric fissure. On the other hand, a "secondary cyst" with an intact falx may be formed by hydrocephalus during the later stage (during the pre- and perinatal period) after falx formation is complete. In 2001, Barkovich et al.[40] reported 15 cases of callosal agenesis with a communicating interhemispheric cyst, which appeared to be an extension or diverticulation of the third or lateral ventricles. These authors also classified the cysts into three types: Type a was associated with

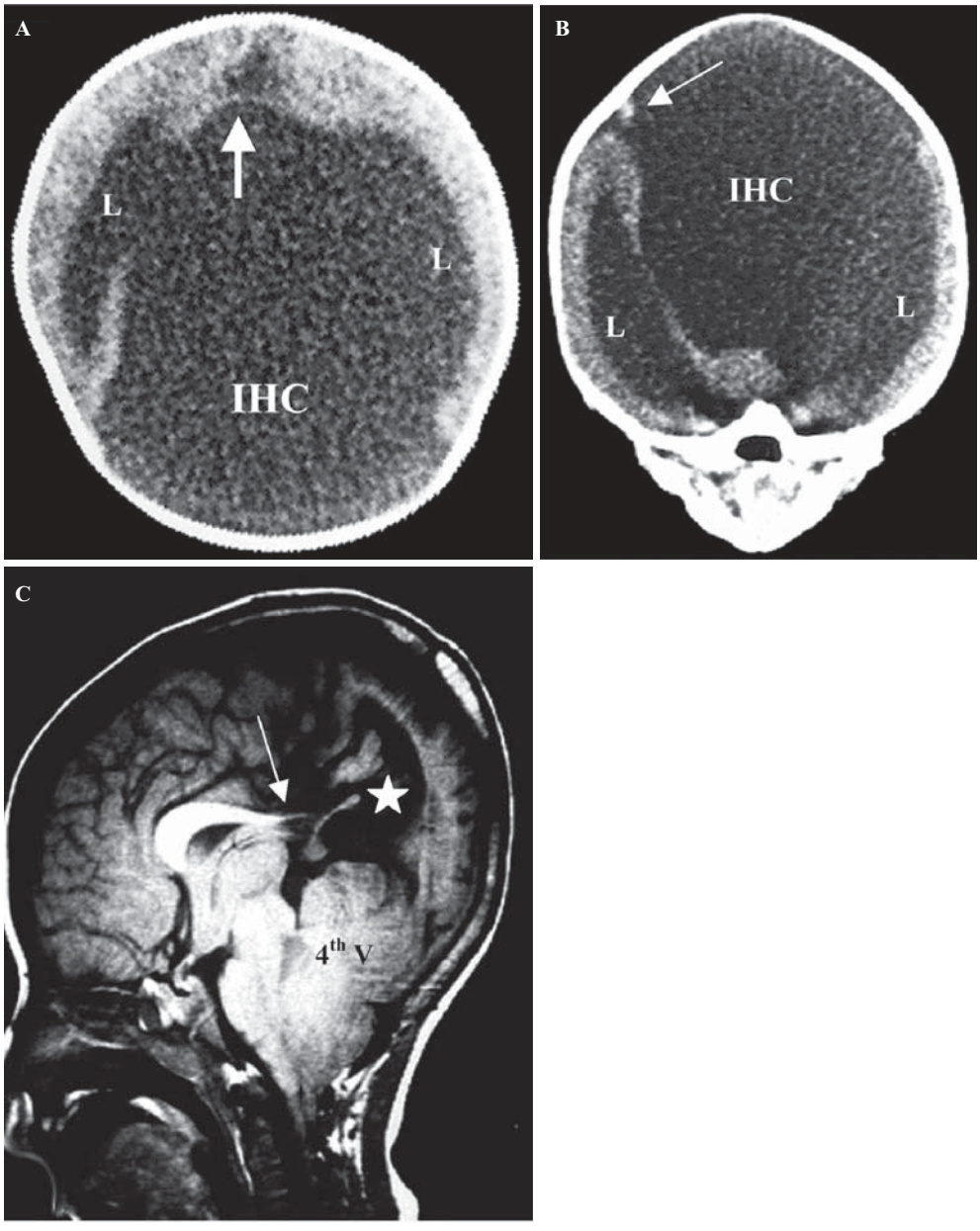


Fig. 11A,B. Primary communicating interhemispheric cyst in a 0-day-old male neonate. Plain CT before a ventriculoperitoneal shunt. Axial image (**A**) shows that a huge interhemispheric cyst (*IHC*), where the anterior wall bulges into the anterior interhemispheric fissure (*arrow*), displaces the body of the lateral ventricles (*L*) bilaterally. Direct coronal image (**B**) shows deficient falx cerebri (*arrow*). **C** T1-weighted midsagittal image at 4 years after the shunt shows a partial defect of the corpus callosum with thinning of the posterior body and a missing splenium (*arrow*). A dilated interhemispheric fissure (*asterisk*), which may have resulted from a previously enlarged third ventricle due to hydrocephalus, is seen. Aqueductal stenosis is also noted. *4th V*, fourth ventricle

presumed communicating hydrocephalus but no other cerebral malformation; type b was associated with hydrocephalus secondary to diencephalic malformations (fused thalami) that prohibited the egress of CSF from the third ventricle into the aqueduct; and type c was associated with a small head size and apparent cerebral hemispheric dysplasia or hypoplasia.

Dorsal cyst with holoprosencephaly

Whereas the hemispheric brain represents normal development of the paired neocortical primordium (telencephalon "totopar"), the holospheric brain, which is an essential feature of holoprosencephaly, results from failed development of the neocortex, which may originate from a single or unpaired primordium. Thus, holoprosencephaly shows an undivided telencephalon "totopar," where the neocortex shows midline continuity in the more rostral part of the telencephalon (Fig. 12).[8,33,41,42] Another common feature of holoprosencephaly is the presence of a dorsal sac membrane, which represents the unfolded diencephalic roof plate and covers the dorsocaudal aspect of the prosencephalic ventricle.[35,41–43]

The term "dorsal cyst" is used to refer to both cystic expansion of the dorsal sac membrane (holospheric dorsal cyst) and an elevated but folded diencephalic roof

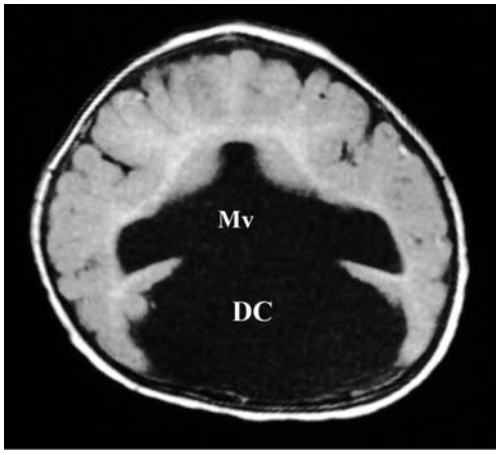


Fig. 12. Semilobar holoprosencephaly with a dorsal cyst in a 1-year-old male infant. Axial T1-weighted image shows the midline continuity of the cerebral cortex without formation of an interhemispheric fissure. Note the dorsal cyst (*DC*), which continues from the monoventricle (*Mv*)

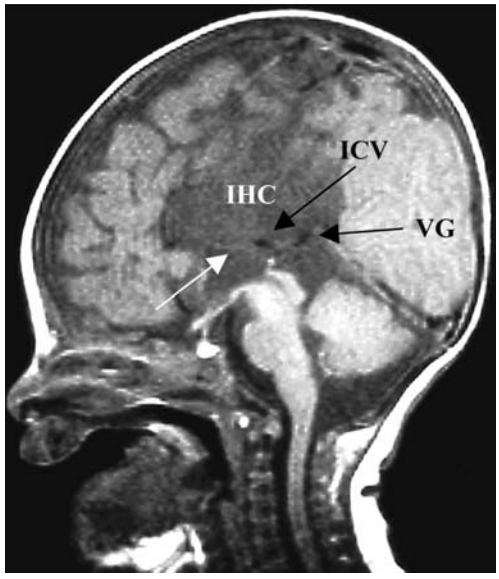
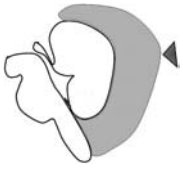
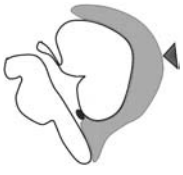


Fig. 13. Noncommunicating interhemispheric cyst in a 27-day-old male infant. Midsagittal T1-weighted image shows absence of the corpus callosum and an interhemispheric cyst (*IHC*) above the roof of the third ventricle (*arrow*). Note that the internal cerebral vein (*ICV*) is displaced inferiorly by the cyst. *VG*, vein of Galen

Table 1. Classification and morphological features of posterior fossa cysts

Parameter	Neuraxial cyst		Extraaxial cyst	
Cyst	Dandy-Walker cyst	Blake's pouch cyst	Arachnoid cyst	Mega cisterna magna
Origin	AMS	AMI	Arachnoid membrane	Arachnoid membrane
Vermis	Agenesis or hypoplasia	Normally formed	Normally formed but displaced anteriorly	Normally formed
Posterior fossa	Large	Normal	Occasionally large	Normal
Dural sinuses	High-positioned torcular with elevated straight and lateral sinus	Normal	Occasionally dysgenetic torcular	Normal
Midsagittal drawing				
Other	Dandy-Walker variant seems to be a Blake's pouch cyst.		Arachnoid pouch Arachnoid cyst without cerebellar compression Cyst extends beyond the IOP Midsagittal drawing 	Cyst must not extend beyond the IOP

AMS, area membranacea superior; AMI, area membranacea inferior; IOP, internal occipital protruberance (triangular symbol in drawing)

Table 2. Classification and morphological features of supratentorial cysts

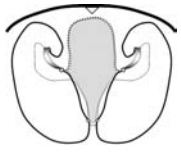
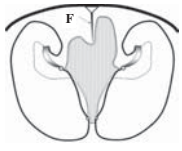
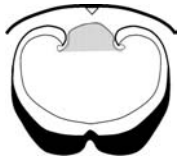
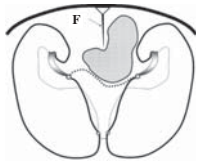
Parameter	Neuraxial cyst			Extraaxial cyst
Cyst	Communicating interhemispheric cyst (hemispheric dorsal cyst)		Dorsal cyst with holoprosencephaly (holospheric dorsal cyst)	Noncommunicating interhemispheric cyst
Origin	Diencephalic roof plate (roof of the third ventricle)		Diencephalic roof plate (roof of the prosencephalic ventricle)	Arachnoid or neuroepithelial membrane
Telencephalon	Hemispheric brain with agenesis of the corpus callosum		Holospheric brain	Hemispheric brain with agenesis of the corpus callosum
Falx/tentorium	Primarys cyst Deficient	Secondary cyst Normal	Deficient	Normal
Galenic system	Dysgenesis	Normal	Dysgenesis	Normal
Coronal drawing				
		F, falx		F, falx
Other	Due to hydrocephalus during the early fetal period	Due to hydrocephalus during the prenatal and neonatal periods	Seen in alobar or semilobar holoprosencephaly	The roof of the third ventricle is displaced inferiorly

plate (hemispheric dorsal cyst). Indeed, a hemispheric dorsal cyst is a communicating interhemispheric cyst with callosal agenesis (Fig. 11).[8,35,42] In fact, whereas a holospheric dorsal cyst in holoprosencephaly represents primary failure of inversion of the unfolded diencephalic roof plate, a hemispheric dorsal cyst (communicating interhemispheric cyst) represents a failure of inversion of the folded diencephalic roof plate secondary to intraluminal pressure in the diocele after the completion of hemispheric cleavage of the telencephalic vesicle. Thus, to differentiate them from one another, it is necessary to distinguish whether the telencephalic vesicle is a holosphere or a hemisphere.

Interhemispheric extraaxial cyst with callosal agenesis

Noncommunicating interhemispheric cyst

Noncommunicating interhemispheric cysts are defined as loculated extraparenchymal cysts that do not communicate with the third or lateral ventricle and are associated with partial or complete callosal agenesis.[39] Pathologically, these cysts consist of an arachnoid membrane (arachnoid cyst) or a neuroepithelial membrane (ependymal cyst). Whereas the roof of the third ventricle of a communicating cyst is elevated superiorly, it is displaced inferiorly by a noncommunicating cyst (Fig. 13). Inaddition, these cysts are usually multiloculated and occasionally accompanied by migration anomalies such as neuronal heterotopia, polymicrogyria, or both.[40]

Conclusions

The classification and morphological features of midline cystic malformation are summarized in Tables 1 and 2. These lesions are often difficult to differentiate on the basis of their radiologic appearance alone. However, an accurate diagnosis is essential for proper treatment planning and genetic counseling. A careful review of neuroembryologic development is necessary to make a more accurate radiologic diagnosis.

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