Absence of the Septum Pellucidum: A Useful Sign in the Diagnosis of Congenital Brain Malformations

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In a review of more than 2000 MR images of the brain we identified 35 patients with absence of the septum pellucidum. These patients were divided into seven basic groups as follows: septooptic dysplasia; schizencephaly; holoprosencephaly; agenesis of the corpus callosum; chronic, severe hydrocephalus; basilar encephaloceles; and porencephaly/hydranencephaly. Absence of the septum pellucidum was never seen as an isolated finding. By using data gathered from the review of the MR scans of patients in this study, we devised a diagnostic algorithm to aid in the classification of these patients.

Absence of the septum pellucidum can provide a valuable clue to the diagnosis of malformations of the brain.

Absence of the septum pellucidum is reported to be an unusual anomaly that occurs in an estimated 2 to 3 individuals per 100,000 people in the general population [1]. When observed, this anomaly should serve as a clue to the presence of associated anomalies, including holoprosencephaly, septooptic dysplasia, abnormalities of the corpus callosum, and Chiari II malformation [1].

The purpose of this study was to identify the underlying brain anomalies in 35 consecutive patients in whom absence of the septum pellucidum was demonstrated by MR imaging, to characterize these malformations, and to develop an algorithmic approach to their diagnosis.

Subjects and Methods

Absence of the septum pellucidum was identified in 35 patients selected from 2007 MR images of the brain obtained over a 2½-year period at the University of California at San Francisco and at the San Francisco Magnetic Resonance Center. Patients' ages ranged from 2 days to 23 years (mean, 2.3 years). Developmental abnormalities occurred as follows: septooptic dysplasia (seven patients), schizencephaly (six patients), Chiari II malformation (seven patients), aqueductal stenosis (five patients), holoprosencephaly (four patients), encephaloceles (three patients—two with associated callosal agenesis), agenesis of the corpus callosum with interhemispheric cyst (two patients), porencephaly (two patients), and hydranencephaly (two patients). Three of the patients with septooptic dysplasia also had schizencephaly. No cases of absence of the septum pellucidum were identified without associated anomalies. All patients were imaged on a 1.5-T GE imager. Imaging parameters included 5-mm thick axial spin-echo (SE) sections with 2500/35,70 (TR/TEs) and sagittal SE images with 600/20 (TR/TE). In some patients, coronal SE 600/20 images were also obtained. The coronal views were especially useful for assessing the interhemispheric fissure in suspected holoprosencephaly and for evaluating the optic chiasm in suspected septooptic dysplasia.

The images were evaluated retrospectively for the presence or absence, size, location, and appearance of the following structures: corpus callosum, fornices, falx cerebri, interhemispheric fissure, lateral ventricles, optic nerves, optic chiasm, cerebral cortex, cerebral white matter, and clefts within the cerebral hemispheres. The results were then compiled and contrasted and an algorithm was devised to facilitate diagnosis (Fig. 1).

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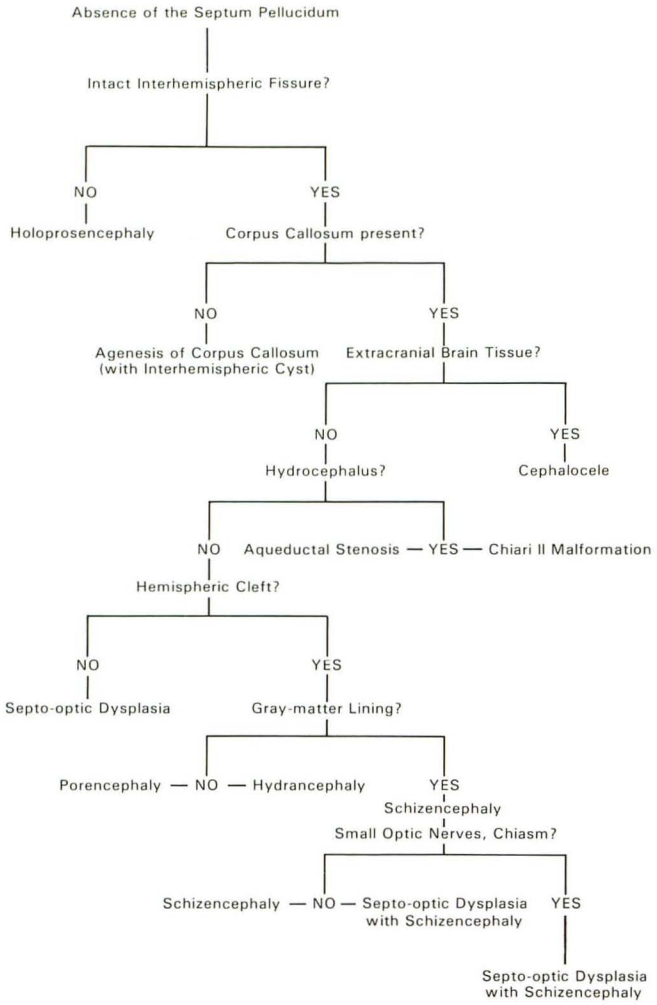


Fig. 1.—Algorithm to facilitate diagnosis of underlying brain anomaly in patients with absence of septum pellucidum.

Results

Septooptic Dysplasia (Seven Patients)

The septum was completely absent in these seven patients. In all of these subjects, the fornix was present but low in location. The fornix was attached to the posteroinferior aspect of the splenium of the corpus callosum (Fig. 2). The corpus callosum was present in all seven patients, but exhibited a focal narrowing in three of the patients with associated schizencephaly. The location of the callosal narrowing correlated with that of the cleft. (One patient with a frontal cleft had a focal narrowing of the genu, two with parietal clefts had narrowing of the callosal body.) This correlation of a callosal narrowing with the location of a hemispheric cleft has been previously described [2]. The falx and interhemispheric fissure were intact and complete. The optic nerves and/or optic chiasm were subjectively small in four of the seven patients. The frontal horns had a square appearance on coronal images in all seven patients; inferior pointing of the frontal horns was identified in three. In the three patients with schizencephaly, the lateral ventricles were of normal size. Of the four patients without schizencephaly, two had enlarged lateral ventricles with diminished white matter in the centrum semiovale and the cortical sulci were shallow. In a third patient the ventricular enlargement was unilateral.

Schizencephaly (Six Patients)

The anterior-most 1 cm of the septum was present in one patient; complete absence was noted in the other five. The fornix was present, but low with posterior displacement of the attachment to the splenium in all six patients. The corpus callosum, falx, and interhemispheric fissure were intact and complete except for focal thinning of the corpus callosum, as described above. The optic nerves and chiasm were small in two of the patients, both of whom also had septooptic dysplasia. In the three patients in whom the frontal horns were not involved by the cleft, the frontal horns had a square appearance. The frontal horns were distorted in the three patients in whom they were involved by clefts (Fig. 3). The cerebral cortex was thickened with an irregular gray matter–white matter junction and shallow sulci along and adjacent to the cleft.

Aqueductal Stenosis (Five Patients)

There was marked hydrocephalus in all five patients. The septum was partially absent in two of the five patients (Fig. 4), nearly completely absent in one, and completely absent in two subjects. In the patients with partial absence of the septum, the middle of the septum was absent but the most anterior, posterior, superior, and inferior portions were present. The septum was progressively thinner close to the absent center (Fig. 4). The fornix was inferiorly displaced in all five cases. The corpus callosum, falx, interhemispheric fissure, optic nerves, and optic tracts showed no developmental abnormality. The corpus callosum was stretched, the frontal horns ballooned, the cortex flattened, and the white matter thinned by the marked hydrocephalus. No other abnormalities were noted.

Chiari II Malformation (Seven Patients)

All seven patients had ventriculoperitoneal shunts for severe hydrocephalus. The septum was completely absent in four patients and absent except for a few remnants in three of the seven. The appearance of the septal remnants at the periphery and of the inferiorly displaced fornices was identical to that of the remnants of septum in the patients with aqueductal stenosis. In all seven patients, the rostrum and splenium of the corpus callosum were absent, and the falx was completely formed in all seven subjects, but was fenestrated in three. The interhemispheric fissure was complete in all seven patients, but in three of the children it was off-center as a result of gyral interdigitations (Fig. 5). The frontal horns of the lateral ventricles were enlarged as a result of hydrocephalus. The trigones and occipital horns were enlarged by

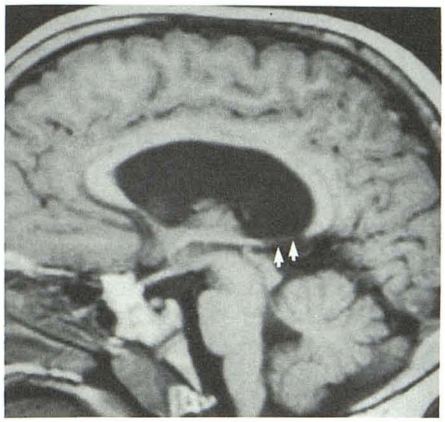


Fig. 2.—Midline sagittal SE 600/20 MR image of patient with septooptic dysplasia. Fornix (*arrows*) is low in position, meeting splenium of corpus callosum posteriorly and inferiorly. Axial images of this patient showed absence of septum pellucidum.

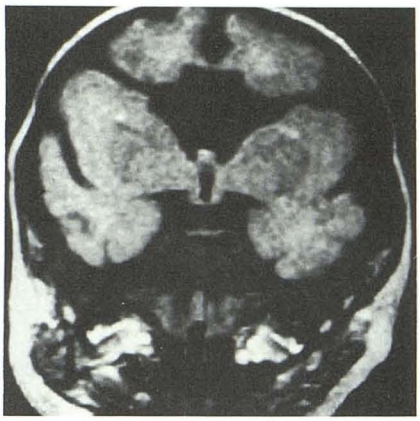


Fig. 3.—Coronal SE 600/20 MR image of patient with bilateral schizencephaly. Septum pellucidum is completely absent. Shape of frontal horns is distorted by clefts.

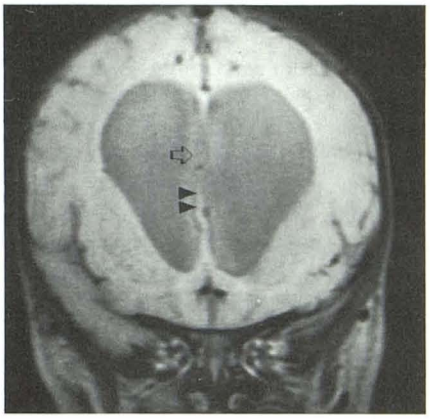


Fig. 4.—Coronal SE 2500/35 MR image of patient with severe hydrocephalus resulting from aqueductal stenosis. Septum is thin inferiorly (*arrowheads*) and absent centrally (*arrow*). A wide range of thinning and partial absence of septum was seen in patients with aqueductal stenosis and Chiari II malformation. Pressure necrosis of septum, resulting from longstanding hydrocephalus, may be the cause of these septal anomalies.

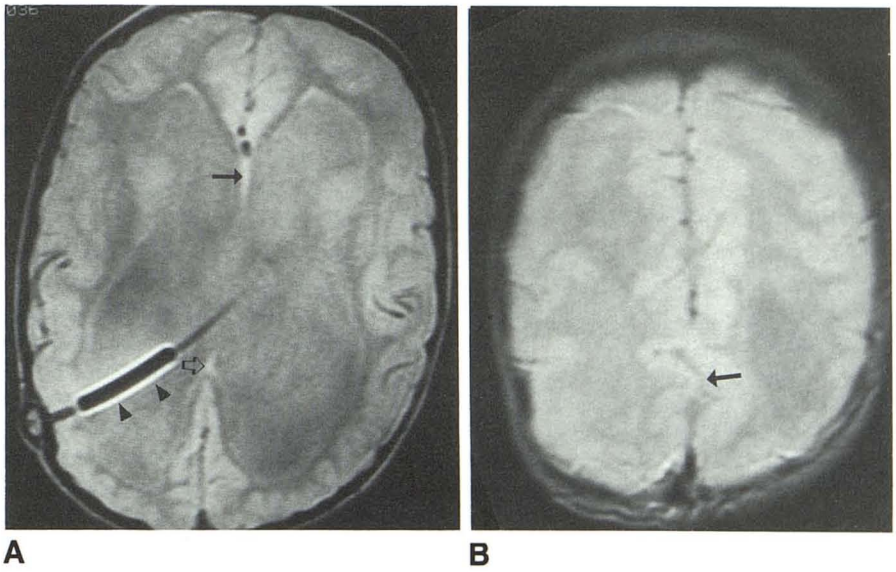


Fig. 5.—Chiari II malformation.

A, Axial SE 2500/35 MR image at level of bodies of lateral ventricles. A ventriculostomy tube is in place in right lateral ventricle (*arrowheads*). Only the most anterior 2 cm (*closed arrow*) and most posterior 1 cm (*open arrow*) of septum are present at this level.

B, Axial SE 2500/35 MR image above lateral ventricles. Gyri of cerebral hemispheres interdigitate through an area of falx fenestration (*arrow*). This is a helpful sign in diagnosing Chiari II malformation.

a combination of chronic hydrocephalus and colpocephaly. The mantle of the cerebral white matter was slightly thinned. The cerebral cortex, optic nerves, and optic chiasm were normal in all seven patients.

Holoprosencephaly (Four Patients)

The septum was completely absent in all four patients who had holoprosencephaly. Three patients had semilobar holoprosencephaly and one had lobar holoprosencephaly. The frontal lobes were entirely fused in two of the infants with semilobar holoprosencephaly. The frontal lobes were fused in the posterior frontal region in the third patient with semilobar holoprosencephaly and in the patient with the lobar variety. None of the four patients had a detectable corpus callosum or fornix. The frontal horns were unformed in the three patients with the semilobar form; they were present, but hypoplastic, in the child with the lobar form. The cerebral cortex and white matter were normal in appearance in the child with lobar holoprosencephaly and in two of the three patients with the semilobar variety. In the third infant with the semilobar form, the cortex was thickened with multiple heterotopias and diminished thickness of the white matter. The optic nerves and chiasm appeared normal in all four patients.

Cephaloceles (Three Patients)

The septum was completely absent in all three patients. In the two patients with frontonasal encephaloceles, there was agenesis of the corpus callosum in one and atrophy of the corpus callosum in the other. In the third patient, with a sphenoidal encephalocele, there was agenesis of the corpus callosum. The fornix was absent in the two patients with callosal agenesis; in the patient with callosal atrophy, the fornix was low with a posteriorly displaced attachment to the splenium. The frontal horns were squared and the ventricles otherwise normal in the patient with an intact corpus callosum; the frontal horns were crescent shaped and the trigones dilated (colpocephaly) in the two patients with agenesis of the corpus callosum. The optic chiasm and intracranial optic nerves were stretched and displaced by the cephaloceles. The interhemispheric fissure and falx cerebri were normal in all three patients. The cerebral cortex and white matter were normal except for stretching near the site of the cephaloceles.

Agenesis of the Corpus Callosum (Two Patients)

In each of two patients who had agenesis of the corpus callosum there was a large associated interhemispheric cyst. The septum and the fornix were completely absent in both patients. The frontal horns were crescentic in shape, and there was colpocephaly in both patients. The interhemispheric fissure was widened by the cyst. The optic nerves and optic chiasm, cerebral cortex, and cerebral white matter were normal.

Porencephaly (Two Patients)

Two patients had CSF-intensity clefts, which were not lined by gray matter, in the cerebral hemispheres. Both patients had complete absence of the septum. The corpus callosum was complete but uniformly thin, and the amount of cerebral white matter was generally reduced in both patients. In one patient, the fornix was absent on the side of the lesion; in the other, the fornix was not well-evaluated. The ipsilateral frontal horns communicated with and were distorted by the clefts. The cerebral cortex, falx, interhemispheric fissure, and optic systems were normal.

Hydranencephaly (Two Patients)

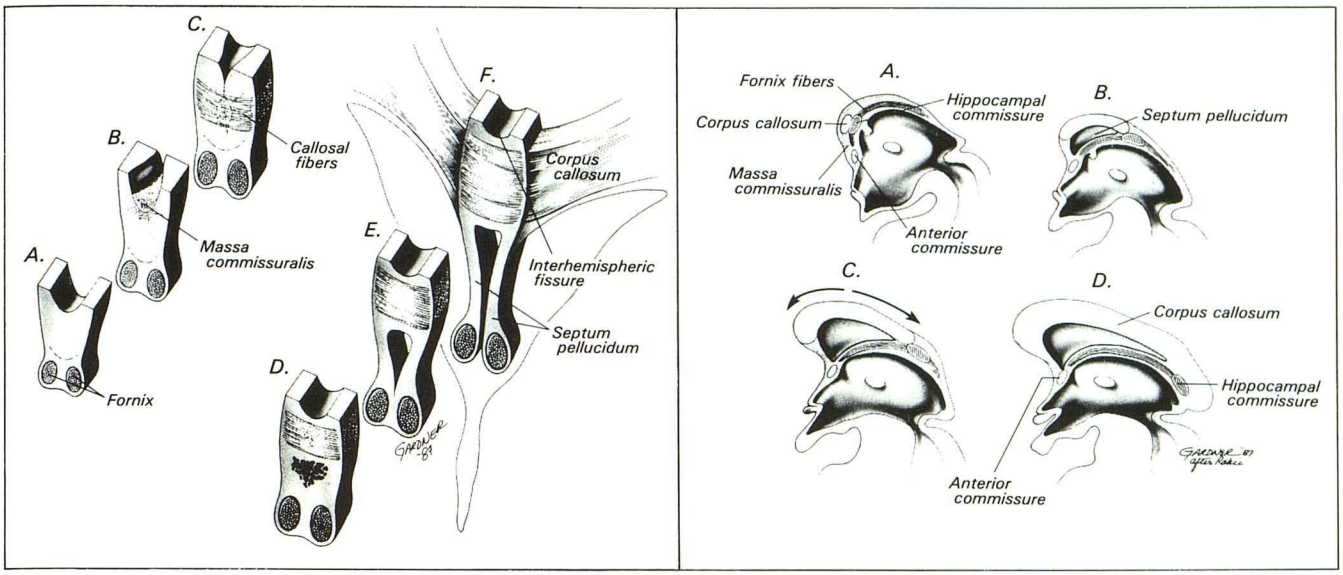
Two patients had complete absence of the hemispheres, except for small foci of remaining tissue in the inferomedial frontal and medial temporal lobes. The septum was completely absent, as were the corpus callosum, fornix, ventricles, interhemispheric fissure, optic systems (except for the optic nerves), and the vast majority of the cerebral cortex and white matter. The falx cerebri was intact in both patients.

Discussion

Proper classification of the dysmorphic brain is most important because of the very different prognoses associated with the different anomalies of the brain. For example, patients with holoprosencephaly are nearly always severely developmentally retarded and rarely live for more than a few years, while those with callosal agenesis can have normal intelligence and lead normal lives. Results of this study indicate that absence of the septum pellucidum can serve as a useful indicator of additional brain malfunction. In our series, associated anomalies included septooptic dysplasia, schizencephaly, holoprosencephaly, agenesis of the corpus callosum, porencephaly/hydranencephaly, basilar encephaloceles, and chronic severe hydrocephalus (Chiari II malformation and aqueductal stenosis). Absence of the septum pellucidum may be reason to develop an algorithmic approach to distinguish among these anomalies (Fig. 1).

The septum telencephali is a term coined by Andy and Stephen [3, 4] to denote the midline structure located in the rostral telencephalon. It is bounded dorsally by the body of the corpus callosum; rostrally by the subcallosal gyrus, hippocampal continuation, and genu of the corpus callosum; rostroventrally by the nucleus accumbens and subcallosal gyrus; and caudoventrally by the anterior commissure, preoptic area, and anterior hypothalamus. It is bounded laterally by the lateral ventricles. Phylogenetically and functionally, the septum has been divided into two components. A thin, membranous, superior portion that contains glial cells and fiber bundles has been called the septum gangliosum [5], but will be referred to in this article as the septum pellucidum as defined by Andy and Stephen. This structure is seen only in higher primates. The caudally located subdivision of the septum telencephali contains well-developed nuclei that correspond with those seen in lower forms of life. This structure is called the septum verum (septum gliosum of Kuhlenbeck) [5]. The septum verum is not clearly differentiated from the subcallosal gyrus or paraterminal gyrus below, nor from the septum pellucidum above. Within the septum verum are the septal nuclei, the nucleus of the diagonal band of Broca, the bed nucleus of the anterior commissure, and the bed nucleus of the stria terminalis [4]. The septum verum contains multiple fiber systems and essentially acts as a central relay station. It is linked with the hippocampus by fornix fibers, with the preoptic and hypothalamic nuclei by the median forebrain bundle, with the amygdaloid nuclei by the stria terminalis, and with the habenula and colliculi by way of the stria medullaris. The diagonal band of Broca runs medially within the ventral septum and relates the limbic system with the olfactory apparatus. In view of the septum verum's function as a regulatory station linking the diencephalon with the limbic system, it is not surprising that its absence may be associated with a rather severe clinical manifestation [1].

The embryological development of the septum pellucidum has not been clearly elucidated. Rakic and Yakovlev [6] have hypothesized that the leaves of the septum form as a result of cavitation of the medial inferior commissural plate during formation of the corpus callosum (Fig. 6A). The commissural plate is a deep midline structure deriving from the primitive lamina terminalis at the rostral end of the neural tube. The anterior commissure, hippocampal commissure, and corpus callosum develop from this structure. Rakic and Yakovlev suggest that after the corpus callosum forms in a certain



A B

Fig. 6.—Schematic drawings illustrate major theories about formation of septum pellucidum.

***A*, Rakic and Yakovlev [6] have suggested that the leaves of the septum form as a result of cavitation of the medial, inferior aspect of commissural plate after corpus callosum has formed from the plate in a certain region. The thin remaining walls of commissural plate in this region become the leaves of the septum while the cavitary center becomes the cavum septi pellucidi.**

***B*, Abbie [7] and Kuhlenbeck [5] contend that the septum forms early in the caudal commissural plate and then undergoes progressive stretching between the anterior commissure, hippocampal commissure, and corpus callosum.**

area, the remaining walls of the commissural plate in this region become the leaves of the septum pellucidum, while the cavitary center becomes the cavum septi pellucidi. The leaves then fuse, in most people, starting posteriorly and progressing rostrally sometime after birth. Others [1, 5, 7] contend that the septum forms early (sixth or seventh week of gestation) in the caudal commissural plate and then undergoes progressive stretching between the anterior commissure, hippocampal commissure, and corpus callosum; the stretching is a result of progressive growth of the corpus callosum and fornix (Fig. 6B). The results of this study are not sufficient to substantiate either theory, although the fact that in one patient with schizencephaly the most anterior 1 cm of the septum was the only part formed suggests formation in an anterior to posterior direction, as suggested by Abbie [7] and Kuhlenbeck [5].

In all patients with absence of the septum pellucidum, the frontal horns, if not distorted by a cleft, had a square appearance. This sign has been described in association with septooptic dysplasia. It appears that in fact the squaring is a consequence of absence of the septum, and is not specific for a single disease. The fornix consistently occupies an abnormally low position in patients with absence of the septum. The caudal displacement of its attachment to the splenium also appears to be a nonspecific consequence of absence of the septum.

The low position of the fornix in these patients presumably results simply from absence of the tethering effect of the septum. When the septum is present, it holds the fornix well above the velum interpositum. When the septum is absent, the bodies of the fornices settle inferiorly onto the velum interpositum. The only other condition in which this low forniceal position is seen is hydrocephalus of the lateral ventricles, in which the septum is stretched and the fornix inferiorly displaced.

The holoprosencephalies are probably the most devastating brain anomalies associated with absence of the septum pellucidum. The basic underlying defect is a lack of induction of the forebrain within the primitive lamina terminalis, which varies in degree [8–10]. The defect in the primitive lamina terminalis results in a lack of formation of the corpus callosum and septum pellucidum. The distinguishing radiographic feature of the holoprosencephalies is a lack of complete separation of the cerebral hemispheres; the hemispheres will be fused in at least one area, usually in the frontal lobes [11]. This fusion was seen only in the patients with holoprosencephaly and in no others. The initial step in the evaluation of patients with absence of the septum pellucidum, therefore, should be examination of the interhemispheric fissure for evidence of fusion of the hemispheres.

Since the corpus callosum and septum pellucidum both derive from the commissural plate and the corpus callosum forms first, absence of the septum in patients with absence of the corpus callosum is consistent with the known embryological development. Absence of the septum may not be readily recognized on cross-sectional imaging exams, because the third ventricle assumes an abnormally high position in the large monoventricle. The high third ventricle is interposed between the lateral ventricles, and the bodies of the lateral ventricles are compressed from the medial aspect by the longitudinal callosal bundles of Probst [12–15] (Fig. 7). A similar appearance may be seen in "agenesis of the corpus callosum with interhemispheric cyst" [16]. In these patients, a large CSF-containing structure continuous with the ventric-

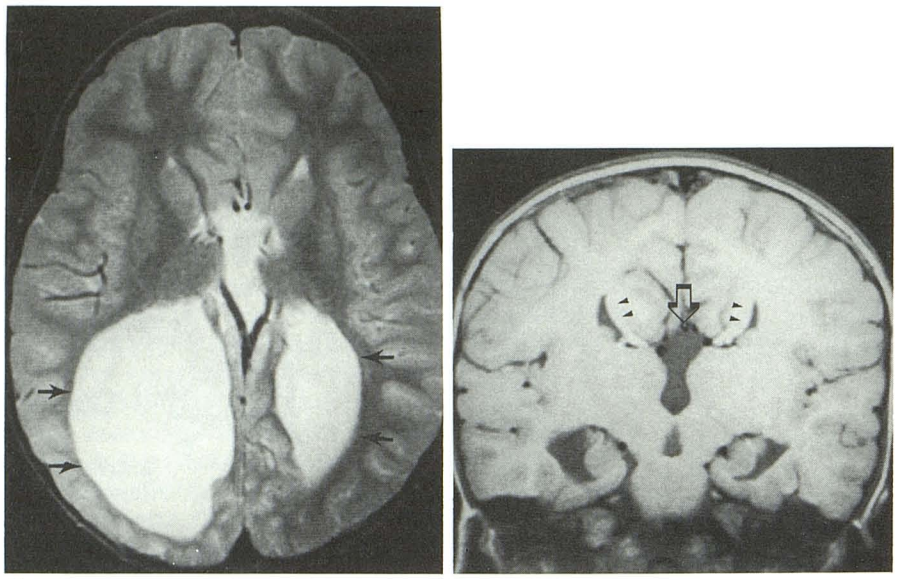


Fig. 7.—Agenesis of corpus callosum.
***A*, Axial SE 2500/70 MR image. Absence of septum pellucidum is not readily apparent because abnormally high-positioned third ventricle is interposed between lateral ventricles. Note enlarged atria and occipital horns of lateral ventricles (colpocephaly), characteristic of agenesis of the corpus callosum (*arrows*).**
***B*, Coronal SE 600/20 MR image reveals abnormally high position of third ventricle (*open arrow*) and longitudinal bundles of Probst (*arrowheads*) impressing the medial aspects of lateral ventricles.**

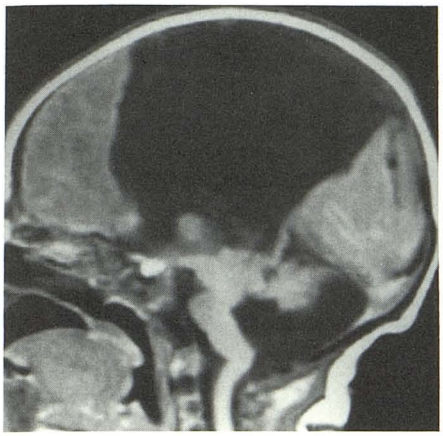
ular system extends upward into the interhemispheric fissure, and the separation between the lateral and third ventricles is lost, giving the appearance of a monoventricle (Fig. 8). Agenesis of the corpus callosum is easily detected on the midsagittal image. The major differentiation is from holoprosencephaly, a diagnosis that has already been eliminated from consideration by identification of a complete interhemispheric fissure, in this case widened by the "cyst."

Two of the three patients with encephaloceles also had agenesis of the corpus callosum, a well-known association in basilar encephaloceles [17]. Basilar encephaloceles may be a result of an abnormality of the primitive lamina terminalis; hence, anomalies of the corpus callosum and septum pellucidum are not surprising. The diagnosis of encephalocele is usually made on clinical grounds. Imaging is done to look for the presence of associated malformations and to determine the location of the vascular structures in the involved region [18]. Since the vast majority of encephaloceles are in the midline, the diagnosis can be made radiographically on the midline sagittal image at the same time the corpus callosum is evaluated. The encephalocele appears as soft tissue extending beyond the calvaria through a calvarial defect. On MR, the low-intensity signal of the calvaria is interrupted at the site of the defect (Fig. 9).

Patients who have Chiari II malformation and patients who have aqueductal stenosis are grouped together in our algorithm, because both groups of patients demonstrate the same spectrum of septal absence. (The patients with the least severe hydrocephalus had a normal septum, and they were not included in this study.) Increasingly large areas of septal fenestration developed as the hydrocephalus became more profound in degree (Fig. 4). In patients with the most severe hydrocephalus, the septum was completely absent. Therefore, the absence of the septum in these patients is presumably the result of septal necrosis from longstanding, severe hydrocephalus. In the present study, all the patients with aqueductal stenosis and Chiari II malformation had severe hydrocephalus and complete or near-complete absence of the septum. The next step in the algorithmic approach, therefore, should be assessment of ventricular size. If there is severe hydrocephalus, sagittal images should be assessed for evidence of tectal beaking; a low, vertical fourth ventricle; a cervicomedullary kink; or herniation of the cerebellum below the foramen magnum. These findings are diagnostic of Chiari II malformation [19–24]. In the absence of these findings, the sylvian aqueduct should be assessed for evidence of narrowing. Diminished flow through the aqueduct can be substantiated on an axial nongated MR image with a long repetition time. Absence of a CSF flow void within the aqueduct supports a diagnosis of aqueductal stenosis [25], although a flow void is absent in a significant number (approximately 10%) of patients without aqueductal disease. A more precise method for evaluating flow through the aqueduct is to do a flow-sensitive, axial, gradient-recalled echo sequence through the level of the aqueduct. A single slice should be obtained by using a flip angle of 50°, TR of 150, and TE of 12.5. Lack of a high signal intensity in the aqueduct supports the diagnosis of a stenotic aqueduct.

In the absence of aqueductal stenosis or Chiari II malformation, the images should be examined for clefts communicating with the lateral ventricles. If no cleft is seen, the patient should be examined for evidence of optic nerve hypoplasia and hypothalamic-pituitary dysfunction. These findings are diagnostic of DeMorsier syndrome (septooptic dysplasia) [26–28]. Examination of the optic nerves is best done clinically, since, in our experience, the optic nerves and chiasm are normal in appearance on MR in about half the patients with DeMorsier syndrome.

When a cleft is present, differentiation should be made between an early developmental or a late destructive lesion. Both the encephaloclastic porencephalies and the developmental schizencephalies are known to be associated with



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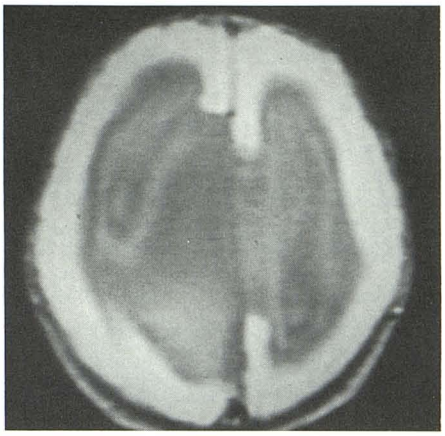

B

Fig. 8.—Agenesis of corpus callosum with interhemispheric cyst.

***A*, Sagittal SE 600/20 MR image. Inferior cerebellar vermis is absent, medulla is kinked, and there is a large fourth ventricle–cisterna magna cyst. These findings are diagnostic of Dandy-Walker malformation. Corpus callosum is absent and CSF extends upward from third ventricle between the cerebral hemispheres to inner table of skull.**

***B*, Axial SE 2500/35 MR image. The appearance is that of a large monoventricle as seen in nearly all patients with absence of septum pellucidum.**

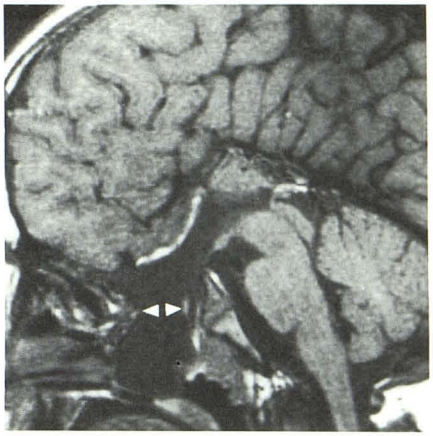

Fig. 9.—Sagittal SE 600/20 MR image of patient with sphenoidal encephalocele. The defect in skull base is seen as a CSF-intensity structure extending through skull base (*arrowheads*). Note associated agenesis of corpus callosum.

absence of the septum pellucidum [1, 29–32]. These entities can be distinguished by the appearance of the cleft. In schizencephaly, the cleft results from a disturbed migration of neuroblasts from the germinal matrix in the subependymal region of the ventricle; this cleft is lined by gray matter [30]. In porencephaly, the cleft results from destruction of tissue after neuroblast migration has begun [32]; gray matter lining of the cleft is absent. Hydranencephaly is classified as a porencephaly; however, this destructive lesion is much more extensive. In hydranencephaly, the entire hemisphere is destroyed, except for areas in the inferomedial frontal and posteromedial temporal regions and the deep central gray matter.

If a gray-matter-lined cleft is present, the optic nerves and chiasm must be examined because of the association between schizencephaly and septooptic dysplasia. As discussed earlier, the clinical examination is the most important factor in this assessment.

In summary, absence of the septum pellucidum is an uncommon finding in imaging studies of the brain. In this series, it was never an isolated finding. Moreover, when this anomaly is identified, it provides a valuable clue to underlying brain malformation. A diagnostic algorithm is provided to facilitate the diagnostic process.

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