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Sonographic Recognition of Agenesis of the Corpus Callosum

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Agenesis of the corpus callosum may be diagnosed successfully in vivo when sonograms demonstrate absence of corpus callosum; absence of pericallosal and cingulate sulci; "sunburst" pattern of sulci along the medial surface of the hemisphere; wide interhemispheric fissure; elevation of the third ventricle; small, laterally positioned frontal horns with concave medial borders; large, laterally positioned and inverted cingulate gyri; and Probst lateral callosal bundles. Comparison of necropsy specimens with sonograms from eight patients with callosal agenesis illustrates the anatomic basis for these diagnostic features.

Agenesis of the corpus callosum (synonym: *callosal agenesis*) designates a group of malformations that range in severity from minor degrees of deficiency of the splenium to total failure of formation of the telencephalic commissures [1, 2]. The malformation may be isolated or form one part of a complex multiple anomaly. Previous authors have described the appearance of callosal agenesis on pneumoencephalograms [3], cerebral angiograms [4], computed tomographic (CT) scans [2, 5-7], and sonograms [8-11]. This report addresses the sonographic features of agenesis of the corpus callosum in greater detail and illustrates the anatomic basis for these features with necropsy specimens.

Materials and Methods

From 1980 to 1983, cranial sonography was performed in eight patients in whom agenesis of the corpus callosum was documented by CT. The eight patients included four girls and four boys who ranged in age from 1 day to 3 months. In each case, sonography was performed in coronal, semiaxial, and sagittal planes through the anterior fontanelle using a Mark III real-time sector scanner (Advanced Technology Labs., Bellevue, WA) with a 5.0 MHz transducer. CT scans were obtained with either an EMI 5005 or GE 9800 scanner. The sonograms from the eight patients were reviewed retrospectively and correlated with necropsy specimens of other patients with callosal agenesis to identify anatomic features that would permit a confident diagnosis of callosal agenesis in vivo.

Normal Anatomy and Sonography

The normal corpus callosum is a thick, crescentic commissure that constitutes 11% of the total volume of the supratentorial brain [1] (fig. 1). It consists of decussating interhemispheric axons that permit learning and memory to be shared between the cerebral hemispheres [12]. The deep surface of the corpus callosum forms the roof of the frontal horns, the roof of the cavum septi pellucidi, and the roof of the bodies of both the lateral ventricles. The superficial surface of the corpus callosum is the pericallosal sulcus and interhemispheric fissure. The paired cingulate gyri course circumferentially around the superficial aspect of the corpus callosum (one cingulate gyrus on each side of the midline) (fig. 1). The cingulate gyri are

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delimited by the pericallosal sulcus on their deep surface, the cingulate sulcus on their superficial surface, and the interhemispheric fissure on their medial surface.

In sagittal section, the sulci on the medial surface of the normal cerebral hemisphere tend to align at an angle to the pericallosal sulcus, rather than truly perpendicular to it (fig. 1).

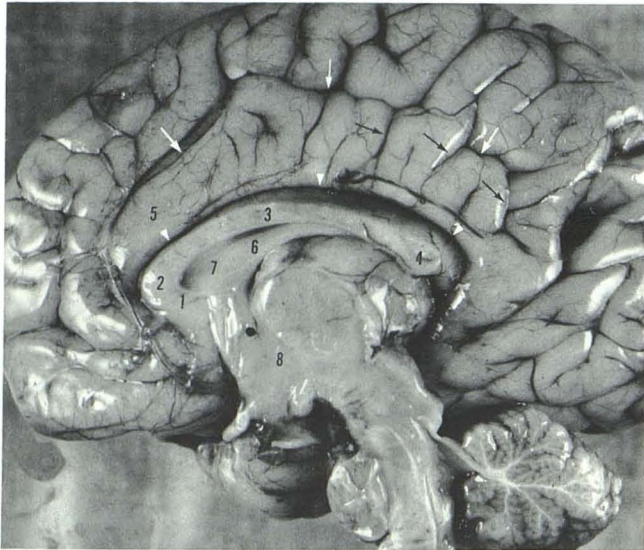


Fig. 1.—Normal. Necropsy specimen. Midsagittal section from 4-day-old boy. Rostrum (1), genu (2), body (3), and splenium (4) of corpus callosum; pericallosal sulcus (arrowheads); cingulate gyrus (5); cingulate sulcus (white arrows); fornix (6); septum pellucidum with cavum septi pellucidum (7); and third ventricle (8). Third ventricle lies well below corpus callosum. Nearly all minor sulci (black arrows) on medial surface of hemisphere terminate at cingulate sulcus, especially frontally; they never extend down to third ventricle.

Nearly all these sulci stop at the cingulate sulcus, well away from the lateral and third ventricle. The third ventricle lies below the lateral ventricle.

On sagittal sonograms (fig. 2), the normal corpus callosum appears as a thin, crescentic hypoechoic band that forms the roof of the frontal horns, cavum septi pellucidum, and ventricular bodies [13, 14]. A thin, hyperechoic line defines the deep surface of the corpus callosum. The normal cingulate gyrus appears as a broader hypoechoic band that is concave with the corpus callosum and lies one level more superficial. The pericallosal sulcus (and its vessels) is seen as a thin, hyperechoic line that separates the corpus callosum from the cingulate gyrus. The cingulate sulcus is seen as a thin, hyperechoic line that delimits the superficial surface of the cingulate gyrus. The sulci on the medial surface of the hemisphere appear as short, thin, hyperechoic lines that lie at an angle to the pericallosal sulcus and that nearly all end at the cingulate sulcus (fig. 2B). The frontal horns, cavum septi pellucidum, and third ventricle are imaged as anechoic spaces deep to the corpus callosum.

On coronal sonograms (fig. 3), the corpus callosum appears as a hypoechoic band that crosses the midline above the cavum septi pellucidum and the two, closely apposed frontal horns. The cingulate gyri appear as paired, paramedian hypoechoic structures that lie above the corpus callosum and are separated from each other by the hyperechoic interhemispheric fissure. The pericallosal sulcus is seen as a thin, V-shaped, hyperechoic line that separates the cingulate gyri from the corpus callosum, while the cingulate sulcus appears as a thin, hyperechoic, inverted V that defines the upper border of the cingulate gyri. The third ventricle is thin and lies well below the frontal horns and bodies of the lateral ventricles.

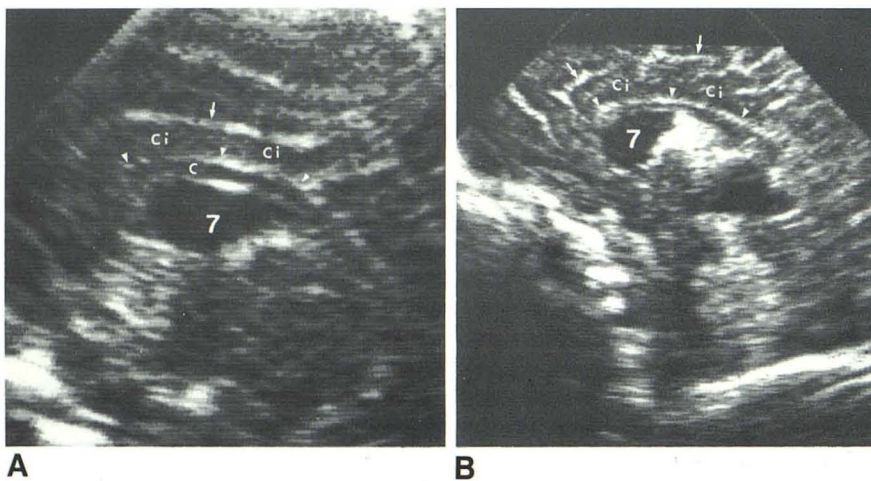


Fig. 2.—Normal midline sagittal sonograms in 3-week-old girl (A) and 4-day-old boy (B). Corpus callosum (C), pericallosal sulcus (arrowheads), cingulate gyrus (Ci), cingulate sulcus (arrows), cavum septi pellucidum (7), and normal arrangement of sulci on medial surface of hemispheres (cf. fig. 1).

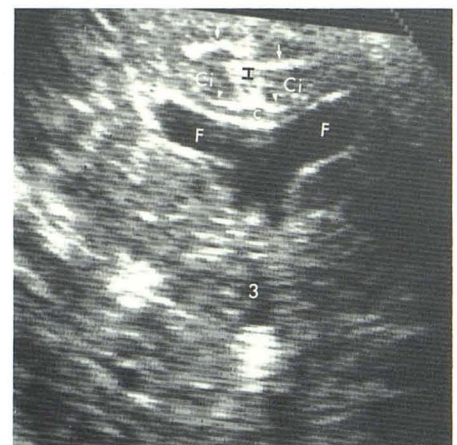
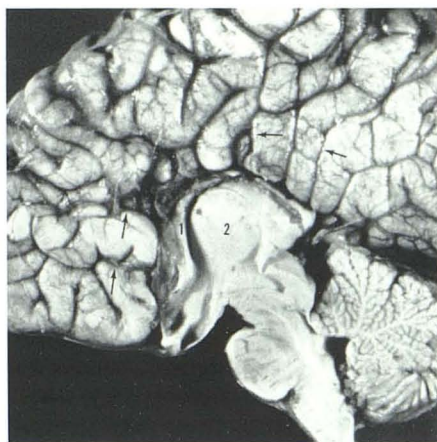


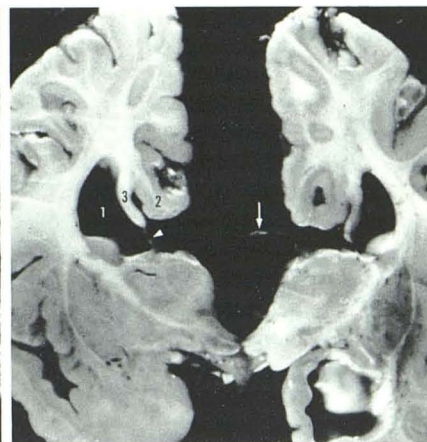
Fig. 3.—Normal coronal sonogram through frontal horns in 2-day-old boy. Thin, smooth interhemispheric fissure (1); paired cingulate sulci (arrows) and pericallosal sulci (arrowheads); corpus callosum (C); and paired cingulate gyri (Ci) situated just next to midline above frontal horns (F). Third ventricle (3) lies well below lateral ventricles.

Fig. 4.—Agenesis of corpus callosum. Necropsy specimen. Midline sagittal section from 3-month-old girl. Complete absence of corpus callosum, pericallosal sulcus, and cingulate sulcus. Fornix (1) and third ventricle (2) ride upward and forward. Sulci (arrows) of medial surface of hemisphere are aligned radial to elevated third ventricle and terminate just superficial to roof of third ventricle. Portion of inferior surface of hemisphere seen in midsagittal plane appears to drape over third ventricle to form narrow arch distinctly different from flatter arch of normal cingulate gyrus in fig. 1.

Fig. 5.—Agenesis of corpus callosum. Necropsy specimen. Coronal section from 2-month-old boy. Absence of corpus callosum; wide interhemispheric fissure; wide, elevated third ventricle (roof membrane [arrow] torn); small, widely separated frontal horns (1) with concave medial margins; and large, laterally positioned, inverted cingula (2) that indent medial surfaces of frontal horns. Medial wall of each frontal horn is formed by cingulum above; Probst white matter bundle (3) in middle; and thin, obliquely inclined sheet of white matter (arrowhead) below.



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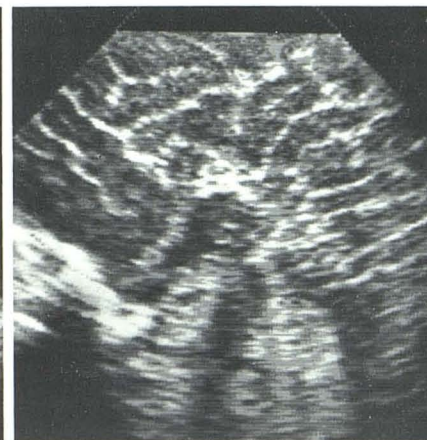


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Fig. 6.—Agenesis of corpus callosum in 4-day-old boy. A, Midline sagittal sonogram. Absence of corpus callosum, absence of pericallosal and cingulate sulci, and high position of third ventricle (arrows). Sulci along medial surface of hemisphere are poorly seen in this plane because interhemispheric fissure is wide. B, Paramedian sagittal sonogram clearly demonstrates radial pattern of sulci along medial surface of hemisphere, convergence of sulci toward roof of elevated third ventricle, and narrow arch of inferior surface of hemisphere (cf. fig. 2).



A



B

Gross Pathology and Sonography of Agenesis of the Corpus Callosum

Complete Agenesis of the Corpus Callosum

In complete agenesis of the corpus callosum, the entire corpus callosum and the normal pericallosal sulcus are absent (fig. 4). The cingulate gyri are laterally positioned, laterally rotated, and often dysplastic in their midportions [2] (figs. 4 and 5). The cingulate sulcus is poorly defined. As a consequence, the cingulate gyri are hard to identify as separate structures on sagittal sections (fig. 4).

The sulci and gyri on the medial surface of the hemisphere have an abnormal orientation directly radial to the roof of the third ventricle (fig. 4). They converge to a narrow, sharply arched inferior margin of the hemisphere [2]. The parietooccipital and calcarine fissures may fail to meet each other [2].

Loss of support from the corpus callosum allows the hemispheres to separate and gives rise to a wide interhemispheric fissure and wide third ventricle (fig. 5). The roof of the third

ventricle is usually intact and, in 80% of cases, extends upward into the interhemispheric fissure to a variable degree [2, 15]. In 52% of cases, the roof of the third ventricle reaches the falx and is indented or deviated by it [2, 15].

In nearly all cases, the axons that would have decussated through the corpus callosum gather into two thick longitudinal bundles of myelinated fibers that course heterotopically from the paraolfactory cortex and frontal lobes to the occipital and temporal lobes. These are the lateral callosal bundles of Probst [2, 15] (fig. 5). The Probst bundles are thickest frontally and thin posteriorly. They lie lateral and usually inferior to the displaced cingulate gyri and are separated from the cingulate gyri by an abnormal cerebrospinal fluid (CSF) space analogous to the lateral ends of the pericallosal sulcus (fig. 5).

The lateral callosal bundles and the cingulate gyri invaginate the medial walls of the lateral ventricles so the ventricular walls are concave medially. The wide separation of the hemispheres and the invagination of the ventricles by the cingulate gyri and Probst bundles cause wide separation of the frontal

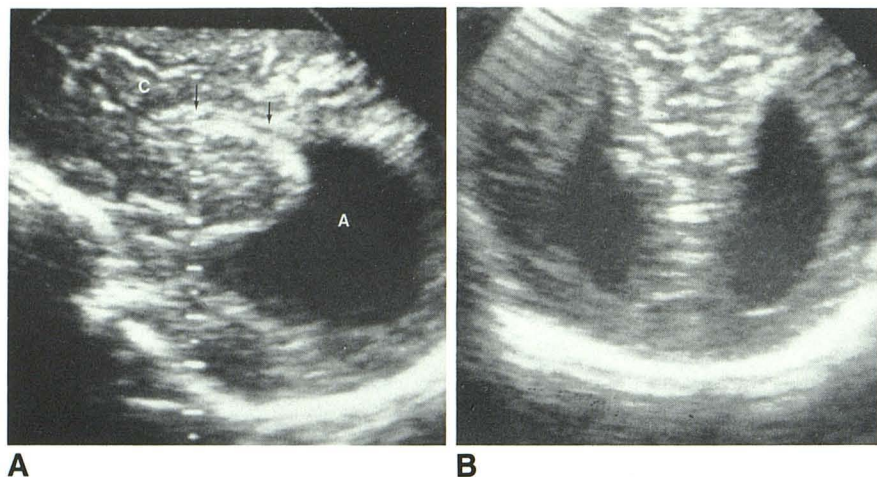


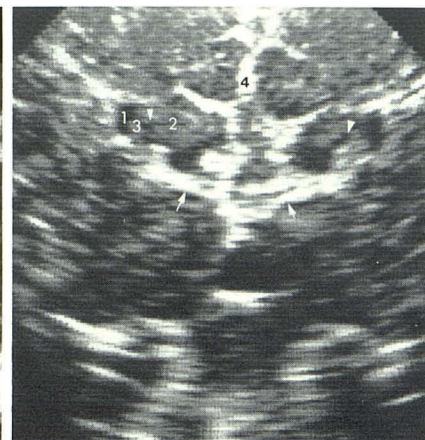
Fig. 7.—Agenesis of corpus callosum in 2-day-old girl. **A**, Lateral sagittal sonogram. Laterally positioned, inverted cingulate gyrus (C), narrow frontal horn and ventricular body (arrows), and substantially dilated atrium (A) and occipital horn. **B**, Semiaxial sonogram. Small, separated frontal horns and dilated atria and occipital horns (colpocephaly). Medial walls of two lateral ventricles diverge anteriorly, forming an angle that is open anteriorly.



Fig. 8.—Agenesis of corpus callosum in 4-day-old girl. Coronal sonogram. Wide, irregular echogenic interhemispheric fissure (1); laterally situated, inverted cingulate gyri (2); and widened, elevated third ventricle (3).



A



B

Fig. 9.—Agenesis of corpus callosum in 2-week-old girl (**A**) and 1-week-old boy (**B**). Coronal sonograms. Small, widely separated frontal horns (1) with concave medial borders; large, inverted cingulate gyri (2); Probst bundles (3); and CSF space (arrowheads) that separates cingula from Probst bundles. Foramina of Monro (arrows) are elongated (cf. fig. 3). 4 = interhemispheric fissure.

horns. The foramina of Monro thus become horizontally oriented and elongated.

The lack of corpus callosum above the ventricular roofs allows the lateral ventricles to extend superiorly into the frontal and parietal white matter [2]. The forceps major is poorly developed. Because the splenium is absent and the forceps major is poorly developed, the atria, occipital horns, and posterior parts of the temporal horns remain dilated in a fetal configuration designated colpocephaly.

Sagittal sonography demonstrates different parts of this deranged anatomy on different sagittal sections. Midline sagittal sonograms demonstrate absence of the corpus callosum and pericallosal sulcus and absence of the cingulate gyri (which have been rotated laterally and no longer lie in or near the midline) (fig. 6A). The high position of the third ventricle is usually evident on this section. Slightly paramedian sagittal

sonograms demonstrate the striking radial ("sunburst") orientation of gyri and sulci on the medial surface of the hemisphere and the narrow arc of the inferior margin of the hemisphere (fig. 6B). More lateral sagittal sonograms demonstrate the narrow, laterally positioned frontal horns and the dilated atria, occipital horns, and posterior temporal horns (fig. 7A). The overall ventricular contour is often best shown on semiaxial sonograms (fig. 7B).

Coronal sonography demonstrates the wide interhemispheric fissure, absence of corpus callosum, high position of the third ventricle (fig. 8), and elongated foramina of Monro (fig. 9). The Probst bundles are best detected by identifying five anatomic features that lie side by side in the same plane: From lateral to medial, these are (1) the small, widely separated, medially concave frontal horns; (2) Probst lateral callosal bundles; (3) the abnormal CSF space analogous to the

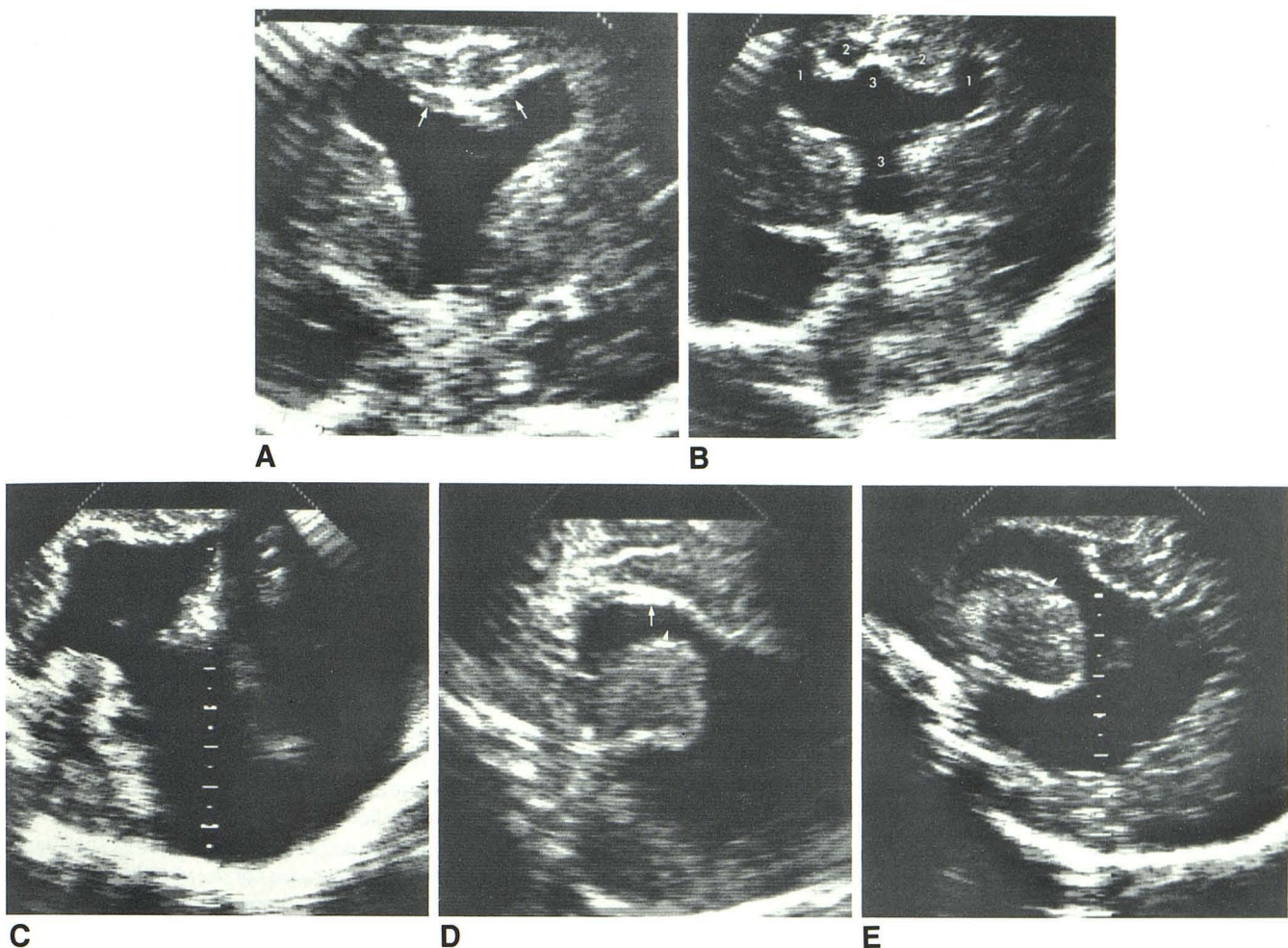


Fig. 10.—Partial agenesis of corpus callosum in 1-month-old girl. **A**, Anterior coronal sonogram at posterior edge of remaining corpus callosum. Cingulate gyri, pericallosal sulcus, and cingulate sulcus are normal. Residual corpus callosum (arrows) shows midline defect. Frontal horns, foramina of Monro, and third ventricle are dilated but otherwise normal. **B**, Slightly more posterior coronal sonogram. Widening of interhemispheric fissure; separated, medially concave lateral ventricles (1); separation of cingulate gyri (2); dilated, elevated third ventricle (3) that bulges upward into interhemispheric fissure; and elongation of posterior aspects of foramina of Monro. **C**, Midline sagittal sonogram.

Large, high-riding third ventricle with preservation of part of corpus callosum anteriorly. Large retrocerebellar space and squat contour of inferior vermis suggest concurrent partial vermian agenesis with Dandy-Walker malformation or variant. **D** and **E**, Paramedian sagittal sonograms demonstrate that corpus callosum (arrow), cingulate gyrus and sulcus, and frontal horn are intact anteriorly. Further posteriorly, ventricle narrows abruptly (arrowheads) just where corpus callosum and cingulate gyrus end and sulci of medial surface of hemisphere radiate down to ventricular margin.

lateral end of the pericallosal sulcus; (4) the large, laterally positioned and rotated cingulate gyrus; and (5) the interhemispheric fissure (fig. 9)

Partial Callosal Agenesis and Interhemispheric Cyst

In partial agenesis of the corpus callosum, a variable segment of corpus callosum is present anteriorly while the posterior segments are deficient. Very rarely, the reverse is true. In such cases, sonography demonstrates the expected normal and pathologic anatomy at different planes of section (fig. 10).

Interhemispheric cysts occur in about 30% of patients with callosal agenesis [5]. Usually they represent marked enlargement of the third ventricle and thus communicate directly with

the third and lateral ventricles. Less often, they appear as multiple loculated cavities of variable histology. In patients with interhemispheric cysts, sonography demonstrates extreme widening of the interhemispheric fissure, compression and inferolateral displacement of the medial surfaces of the hemispheres, inferolateral displacement of the lateral ventricles, (usually) asymmetric bulging of the calvaria with paramedian insertion of the falx, flattening of the cyst against the falx, invagination of the cyst by the falx, and any intervening septa of a multilocular cyst (fig. 11).

Concurrent Malformations

Although agenesis of the corpus callosum may exist as an isolated phenomenon, 80% of cases have other central nerv-

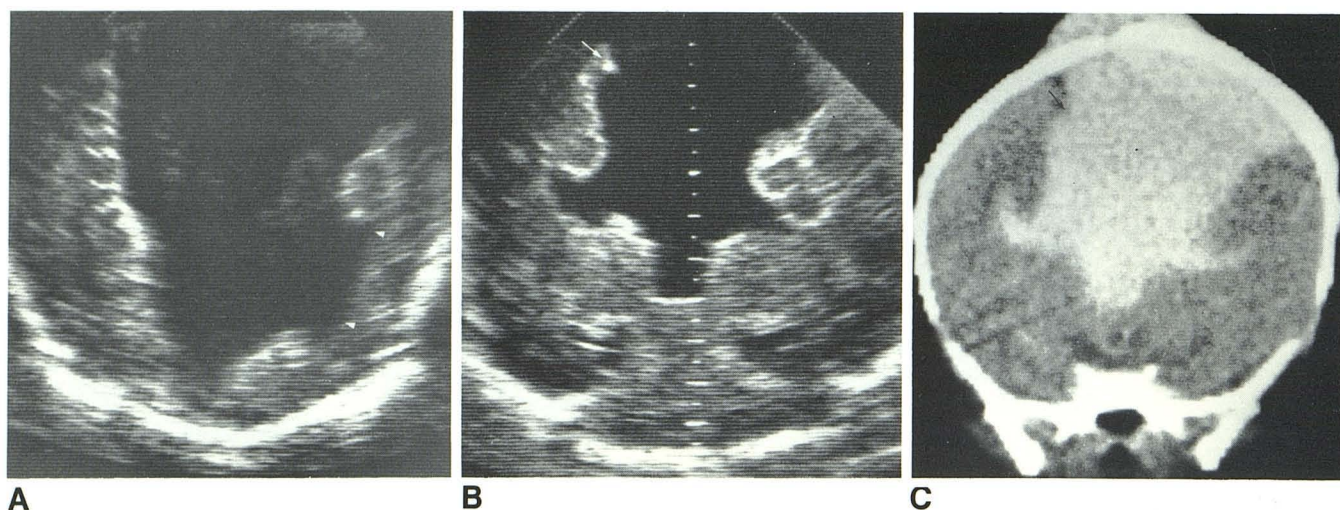


Fig. 11.—Agenesis of corpus callosum with interhemispheric cyst in 4-day-old girl. **A**, Semi-axial sonogram. Large midline cyst displaces gyri and sulci of left hemisphere laterally and communicates with medial aspect of right lateral ventricle (*arrowheads*). **B**, Coronal sonogram. Interhemispheric cyst communicates directly with both lateral ventricles and third ventricle. Cyst exhibits a flat lateral border where it abuts falx and a notch (*arrow*) where it extends under free edge of falx to other side. Callosal agenesis is evident as small, widely

separated frontal horns with concave medial borders; large, inverted cingula; and wide, elevated third ventricle in direct continuity with interhemispheric cyst. **C**, Direct coronal CT scan after instillation of metrizamide via puncture of cyst at anterior fontanelle confirms direct communication of cyst with entire ventricular system, notching (*arrow*) of border of cyst by free edge of falx, outward rolling and displacement of hemispheres by the cyst, and changes of callosal agenesis.

ous system anomalies [6, 16] including interhemispheric cyst (30%) [5]; Dandy-Walker malformation (17%) [17]; and Chiari II malformation, lipoma, and porencephaly [5]. Parrish et al. [16] reviewed the non-nervous system malformations associated with callosal agenesis in 47 autopsy cases.

Discussion

Callosal agenesis is a common anomaly. It has been observed on 0.7% of 2400 cranial CT scans at a children's hospital [5] and on 2.4% of pneumoencephalograms and autopsies in mentally retarded patients [17]. The defect may be asymptomatic. More often it manifests clinically in one or more ways: seizures, mental retardation, microcranium or macrocranium, or symptoms of diverse concurrent central nervous system and distant anomalies [1–16].

Embryologically, the corpus callosum is the largest of three forebrain commissures that originate from the midline of the rostral end of the neural tube near the area of closure of the anterior neuropore. This closure occurs at about 24 days gestational age. Soon thereafter, commissural structures start developing within this area in the primitive lamina terminalis. The active part of the lamina terminalis, called the lamina reuniens, thickens and forms the commissural plate. This plate provides a pathway through which axons may cross from hemisphere to hemisphere.

Within this commissural plate, the forebrain commissures develop. The anterior commissure forms the ventral part of the commissural plate at about 50 days gestational age. The earliest fibers of the corpus callosum appear between the anterior and hippocampal commissures at 10–12 weeks gestational age. The adult form of the structure is achieved by 17 weeks' gestational age [12], but the corpus callosum

continues to increase in thickness as the cortex matures [2].

The precise localization of developing callosal fibers remains somewhat controversial. The most accepted theory states that the earliest fibers lie anterior and inferior to the foramina of Monro. Subsequent fibers of the corpus callosum develop first superiorly and then in a caudal direction, resulting in the characteristic crescentic appearance of the corpus callosum as seen in the sagittal plane [1]. Disruption of this posterocaudal formation could then result in defective development of the splenium. This has indeed been observed often in reports of partial agenesis of the corpus callosum. However, other reports of isolated defects of the anteriorly situated genu of the corpus callosum despite an intact body and splenium suggest the possibility of either an alternative developmental sequence, such as convergence from a multifocal origin [18, 19], or focal destruction of part of a previously well formed corpus callosum.

The most common condition that mimics agenesis of the corpus callosum is shunted hydrocephalus [20, 21]. Severe hydrocephalus thins the corpus callosum remarkably. After shunting, the lateral ventricles collapse and elongate. The thalami move upward, anteriorly and medially, compressing the frontal horns. The interhemispheric fissure "pouts open." The thin, intact corpus callosum drops inferiorly "into" the ventricle and becomes hard to detect. These changes can closely mimic the laterally separated, medially concave frontal horns; elongated foramina of Monro; and high interposed third ventricle—a "pseudoagenesis."

Semilobar and lobar holoprosencephaly can also be confused with agenesis of the corpus callosum [2, 22]. There are several differentiating characteristics: (1) separated thalami in agenesis of the corpus callosum versus fused thalami in holoprosencephaly; (2) presence of Probst bundle in agenesis

of corpus callosum; and (3) presence of fornices in agenesis of corpus callosum.

Occasionally, other midline cavities can simulate an interposed third ventricle. These include cavum septi pellucidi, cavum vergae, and cavum veli interpositi; interhemispheric arachnoid cysts not associated with callosal agenesis; and medial porencephalic cysts. Usually, these conditions can be distinguished from agenesis of the corpus callosum by careful identification or exclusion of the pathologic features of agenesis outlined in this report.

In summary, the frequent use of sonography as a screening tool in neonates and infants has increased the possibility of detecting central nervous system anomalies such as agenesis of the corpus callosum. Familiarity with the pathologic features of callosal agenesis illustrated in this report should permit correct diagnosis in nearly all cases. It must be remembered, however, that accurate diagnosis of callosal agenesis rests on the combination of the many features discussed, rather than on identification of a single "key" sign.

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