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MR Imaging of Callosal and Corticocallosal Dysgenesis

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Callosal dysgenesis implies a malformation of the corpus callosum with origins in the embryogenesis of the telencephalon. A retrospective review of 15 cases of callosal dysgenesis revealed three distinct categories: agenesis (three subjects), hypogenesis (nine subjects), and hypoplasia (three subjects). The basis of this distinction rests upon considerations of neural tube closure, formation and maintenance of the inductive plate of the massa commissuralis, and migration of the neuronal elements of the cerebral cortex responsible for the projection of the commissural fibers of the corpus callosum. These processes are somewhat interdependent in their expression and consequence, resulting in a unique mosaic of callosal and cortical gray matter and hemispheric white matter configurations that have individually varying clinicoradiologic manifestations.

The direct multiplanar imaging capabilities of MR lend themselves ideally to the evaluation of callosal dysgenesis together with the important associated hemispheric findings. Older classification systems, including simply agenesis and partial agenesis, were found to be inadequate for a precise description of the spectrum of developmental callosal anomalies. MR illustrates a range of patterns that allows a categorization of morphologic forms of callosal dysgenesis on the embryogenesis of the telencephalon.

Materials and Methods

The study consisted of a retrospective review of 15 subjects with callosal dysgenesis. T1-weighted, single-echo imaging, 500/40/4 (TR/TE/excitations), was performed in the sagittal and coronal planes with 5-mm-thick sections on a Diasonics 0.35-T MR unit. In addition, a T2-weighted, double-echo acquisition, 3000/40-80/2, was obtained in the axial plane. Patterns were sought in the morphology of the corpus callosum and the cerebral hemispheres, which revealed a predictable relation to embryonic development, both in regard to the decussation of the corpus callosum and to the formation of the cortex responsible for projection of the callosal commissural fibers. No Chiari-type malformations were included in this study group, since the series was limited to CNS anomalies primarily located within the cranium.

Results

MR enabled the differentiation of three distinct categories of dysgenesis (Table 1). In initiative *agenesis* (Type I), the corpus callosum was completely absent (three subjects, Figs. 1 and 2). In *hypogenesis* (Type II), the corpus callosum was variably curtailed caudally in its development owing to primary interruptive factors (five subjects, Fig. 3), or factors related to organic obstruction (four subjects, Fig. 4). This latter subcategory of obstructive hypogenesis was observed solely in cases of so-called "lipoma" of the corpus callosum. In *hypoplasia* (Type III), the corpus callosum was completely formed although focally or generally small in size and associated with prominent dysgenesis of the hemispheric cerebral cortex (three subjects, Figs. 5 and 6). The cortical dysgenesis encountered in this series included

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examples of polymicrogyria, pachygyria, schizencephaly, gray matter heterotopias, and hemimegalencephaly.

An additional finding was the relatively high association of colpocephaly together with the primary callosal abnormality. This included two subjects with agenesis and four with hypogenesis of the corpus callosum (Fig. 2). Other concomitant cerebral observations were a posterior fossa Dandy-Walker midline cyst (one subject) and hypoplasia of the cerebral peduncle(s) (three subjects, Table 2).

Discussion

The embryogenesis of the corpus callosum begins with the formation of the massa commissuralis between the laminae reunions in the dorsal aspect of the lamina terminalis at 10–12 weeks fetal gestation. This structure was initially described in rat embryos by Zuckerkandl in 1901 and confirmed in

human fetuses by Rakic and Yakolev in 1968 [1]. The massa commissuralis acts as the induction bed or plate for the decussation of callosal commissural fibers. Quite simply, if the massa commissuralis does not form, no callosal fibers decussate, and primary or “initiative” agenesis of the corpus callosum results.

However, initial induction is only the first step, and thereafter this induction bed must be maintained throughout decussation. If an arrest of this process occurs at any time during the formation of the corpus callosum spanning 12 to 20 weeks gestation, “interruptive” hypogenesis results. This is the primary cause of terminal hypogenesis of the corpus callosum, which is manifested radiologically by individually varying degrees of caudal curtailment [2]. The etiology of both primary initiative agenesis and interruptive hypogenesis as well as other congenital cerebral malformations are unknown, although infectious agents, radiation, chemical agents, maternal hormones, nutritional deficiencies, hypoxia, and chromosomal and genetic factors have all been considered [3].

A second form of hypogenesis of the corpus callosum has an organic obstruction as its origin [4]. In the cases in this series, the only form of obstruction encountered was that of various-sized lipomatous masses. The explanation of this phenomenon seems to lie with neural tube closure. This closure occurs at 3 to 5 weeks gestation and thus considerably antedates decussation of the corpus callosum between 12 and 20 weeks [1, 5, 6]. It is during this time of neural tube formation that heterotopic rests of tissue may be included within the depths of the developing telencephalon. One of the more commonly encountered inclusions is the lipoma [4, 6]. Once this inclusion mass is placed, if deep and in the midline, it may be in a position that will subsequently result in a head-on collision with the developing corpus callosum, growing

TABLE 1: Categories of Callosal Dysgenesis

Prior Classification	New Classification
I. Agenesis	I. Agenesis
	A. Initiative (\pm cortical dysgenesis/ Pb hypoplasia)
	B. Obstructive (\pm cortical dysgenesis/ Pb hypoplasia)
II. Partial agenesis	II. Hypogenesis
	A. Interruptive (\pm cortical dysgenesis/cc hypoplasia)
	B. Obstructive (\pm cortical dysgenesis/cc hypoplasia)
III. None	III. Hypoplasia ($++$ cortical dysgenesis)

Note.—Pb = Probst bundle; cc = corpus callosum.

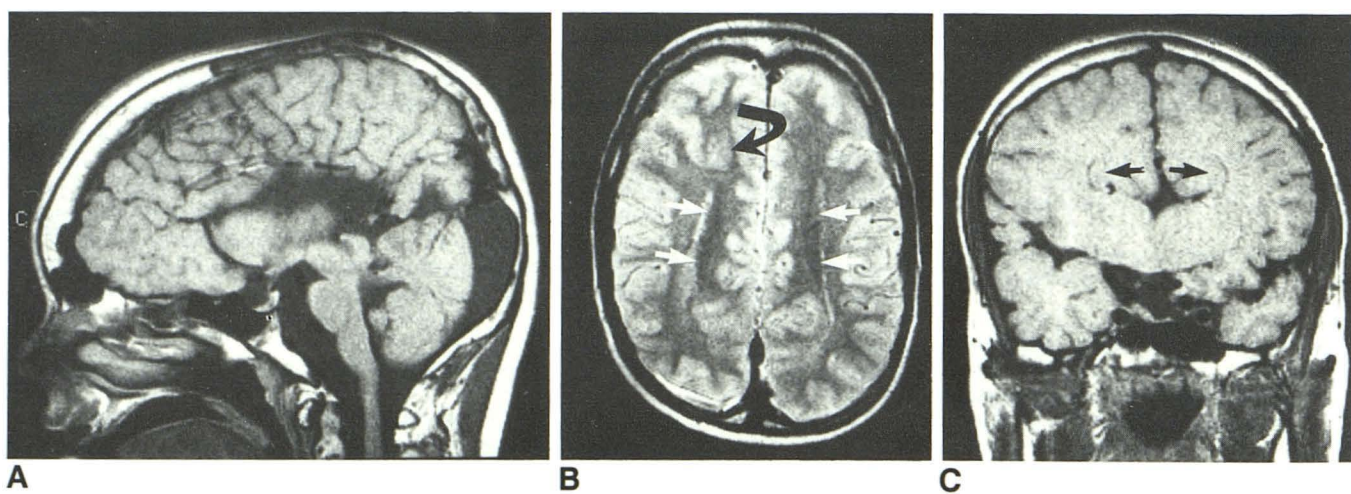


Fig. 1.—Case 3.

A, Sagittal section illustrates totally absent corpus callosum, radiating posterior parietooccipital gyri, and hypoplastic brainstem (confirmed on axial sections).

B, Axial section demonstrates hypointense bands traversing medial borders of lateral ventricles, representing prominent Probst bundles (straight arrows). Also noted is isolated area of gray matter heterotopia on the right frontally (curved arrow).

C, Coronal image shows plump configuration of Probst bundles bilaterally (arrows), reflecting their relative completeness.

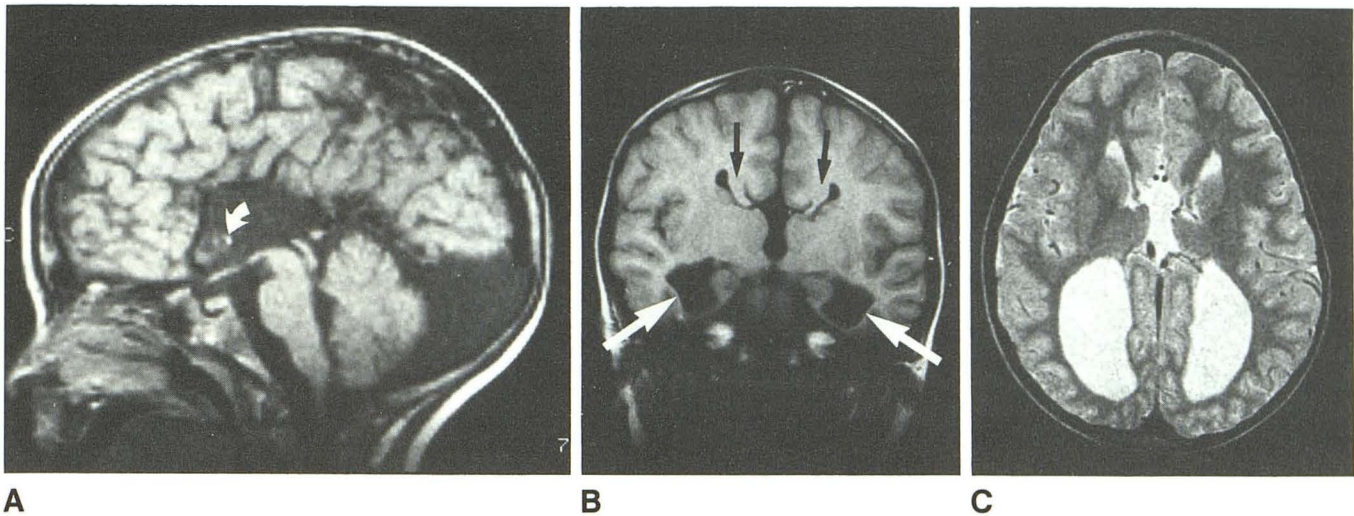


Fig. 2.—Case 2.

A, Sagittal section demonstrates corpus callosum agenesis and intact anterior commissure (arrow).

B, Coronal section illustrates thinned mildly hypoplastic Probst bundles along medial borders of lateral ventricles (black arrows) and colpocephaly involving the temporal horns of the lateral ventricles (white arrows).

C, Axial section through mid-thalamic region shows prominent colpocephaly indicated by dilated occipital horns and trigones of lateral ventricles, likely reflecting the presence of overlying radiologically occult polymicrogyria.

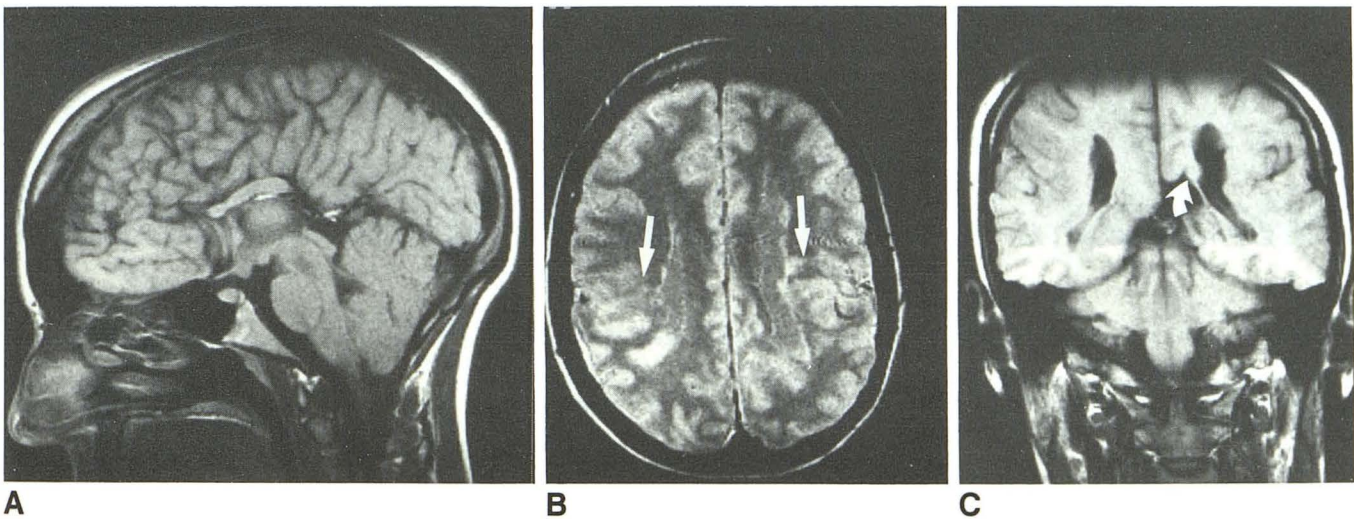


Fig. 3.—Case 7.

A, Sagittal section illustrates a case of primary interruptive hypogenesis of the corpus callosum revealed by the absence of the posterior body and splenium.

B, Axial section through bodies of lateral ventricles demonstrates bilateral gray matter heterotopias (arrows).

C, Coronal slice through trigones of lateral ventricles shows segmental Probst bundle on left (arrow) made more obvious by the focal overlying temporooccipital cortical tissue loss/hypoplasia.

caudally (Fig. 7). This embryonic collision results in the typical cometlike lipomatous callosal cloaking frequently observed (Fig. 4). The degree of secondary hypogenesis associated with these inclusions varies with several factors, including the size of the obstructing mass and its embryonic placement both rostocaudally and ventrodorsally. In fact, complete agenesis might conceivably result if the inclusion mass were located far rostrally between the two halves of the invaginating laminae reunions, thus preventing their apposition and causing an obstructive agenesis [1]. This explains the asso-

ciation of complete agenesis and lipoma reported in the literature although not seen in this series of subjects. Of course, there is also the possibility of an inclusion mass coexisting incidentally with primary agenesis of the corpus callosum and therefore having no obvious direct role in the generation of the callosal malformation. Quite probably, however, many previously reported cases of callosal lipoma may have actually represented varying degrees of obstructive hypogenesis rather than absolute agenesis. While it seems possible that other types of ectopic inclusions might cause

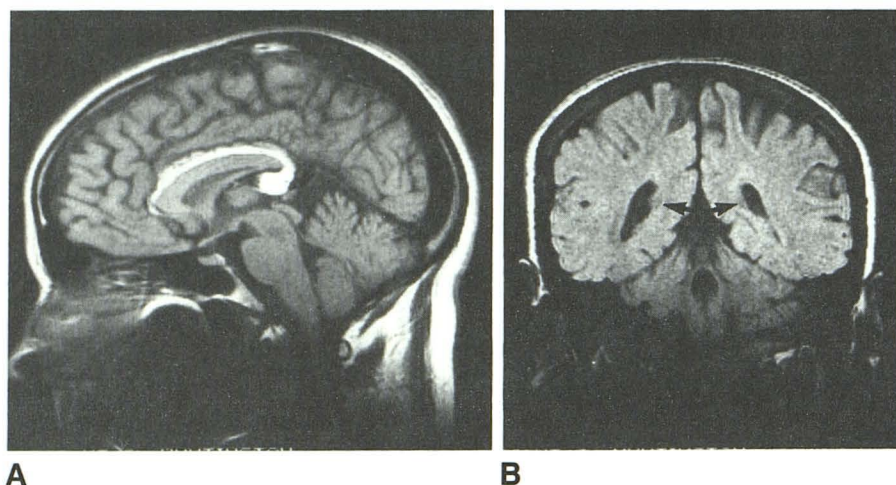


Fig. 4.—Case 10.

A, Sagittal section demonstrates cometlike lipomatous cloaking of corpus callosum with terminal minimal obstructive hypogenesis of splenium.

B, Coronal section illustrates diminutive segmental Probst bundles traversing medial aspect of atria of lateral ventricles (arrows).

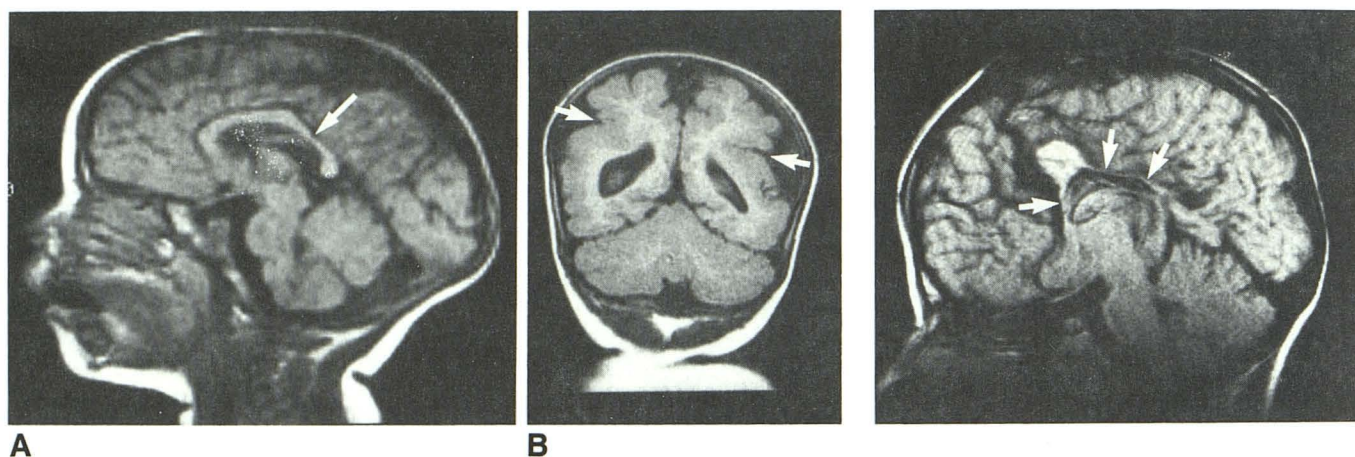


Fig. 5.—Case 13.

A, Sagittal section illustrates focal hypoplasia involving posterior body of corpus callosum (arrow).

B, Coronal section demonstrates incomplete bilateral schizencephaly (arrows) and associated pachygyria within cerebral clefts. These findings were localized on MR to the posterior parietal regions, thus corresponding to the focal hypoplasia in A.

Fig. 6.—Case 15.

Sagittal section illustrates multifocal hypoplasia of corpus callosum involving the genu as well as the posterior body and splenium (arrows). This largely correlates with mosaic polymicrogyric cortex of medial hemispheric surface(s) in low frontal, posterior parietal, and occipital regions.

varying degrees of obstructive hypogenesis (i.e., epidermoid, dermoid, teratoma), none was observed in the present study.

An important ancillary finding in both agenesis and hypogenesis was that of Probst bundles. These structures are peculiar to these conditions and represent the uncrossed callosal fibers running rostrocaudally, parallel to the interhemispheric fissure in the medial walls of the lateral ventricles [1, 4]. These fibers apparently have no neurologically meaningful terminations. Barring associated anomalies, these bundles are complete in their hemispheric representation in agenesis of the corpus callosum (Fig. 1) and segmental in the cases of hypogenesis (Figs. 3 and 4). If concomitant dysplasias are present within the gray and/or white matter of the cerebral hemispheres, Probst bundles may also be hypoplastic, reflecting a reduced number of commissural fibers projecting from the dysgenetic cerebrum (Fig. 2) [7].

The third major category of callosal dysgenesis in this series was hypoplasia. Strictly defined, hypoplasia indicates a struc-

ture that contains all its basic components but that does not reach adult size. Any abnormality in neuronal migration or lamination of the cortical layers will result in cortical dysgenesis [7–11]. Since the cell bodies responsible for the callosal fibers lie within the cerebral cortex, a dysgenetic cerebral cortex will simply not project the “normal” complement of axons to the midline for decussation. As these abnormalities in genesis of the cortex are somewhat random, often incomplete, and not necessarily symmetric, the corpus callosum may consequently demonstrate varying degrees of unifocal, multifocal, or generalized corticogenic projectional hypoplasia (Figs. 5 and 6). As in callosal dysgenesis, the nature of these dysgenetic changes within the cerebral cortex leading to hypoplasia of the corpus callosum is nonspecific and unknown.

The high incidence of colpocephaly (40%) in this series would seem to indicate a possible relationship between callosal, cortical gray matter, and extracallosal cerebral white

TABLE 2: Summary of Patients with Callosal Dysgenesis

Age (years)	Gender	Clinical History	Corpus Callosum Dysgenesis	Neural Tube Inclusion	Associated Gray Matter Dysgenesis	Associated White Matter Dysgenesis	Other Abnormality
4	M	DD	Agenesis	—	—	Colpocephaly	—
5	M	MR	Agenesis	—	—	Colpocephaly	—
24	M	MR	Agenesis	—	—	—	L-HCP
½	M	DD	Hypogenesis	—	Parietooccipital polymicrogyria	Colpocephaly	—
1	M	DD	Hypogenesis	—	Generalized polymicrogyria	Colpocephaly	Bilateral HCP
23	F	MR	Hypogenesis	—	—	Colpocephaly	Dandy-Walker malformation
28	F	MR	Hypogenesis	—	Bilateral gray matter heterotopia	—	—
61	M	MR	Hypogenesis	—	Occipital polymicrogyria	Colpocephaly	—
35	M	Headache	Hypogenesis	Lipoma	—	—	—
38	F	Depression	Hypogenesis	Lipoma	—	—	—
59	F	L-hemiplegia	Hypogenesis	Lipoma	—	—	Infarct, R-internal capsule
59	M	Dementia	Hypogenesis	Lipoma	—	—	Cerebral atrophy, communicating hydrocephalus
1	M	DD, SZ	Focal hypoplasia	—	Focal pachygyria, schizencephaly	—	—
8	M	DD, SZ	Generalized hypoplasia	—	L-hemimegalencephaly, L-gray matter heterotopia	L-hemimegalencephaly	L-HCP
10	M	MR, SZ	Multifocal hypoplasia	—	Mosaic polymicrogyria	—	—

Note.—L = left; R = right; DD = developmental delay; MR = mental retardation; SZ = seizure; HCP = hypoplastic cerebral peduncle.

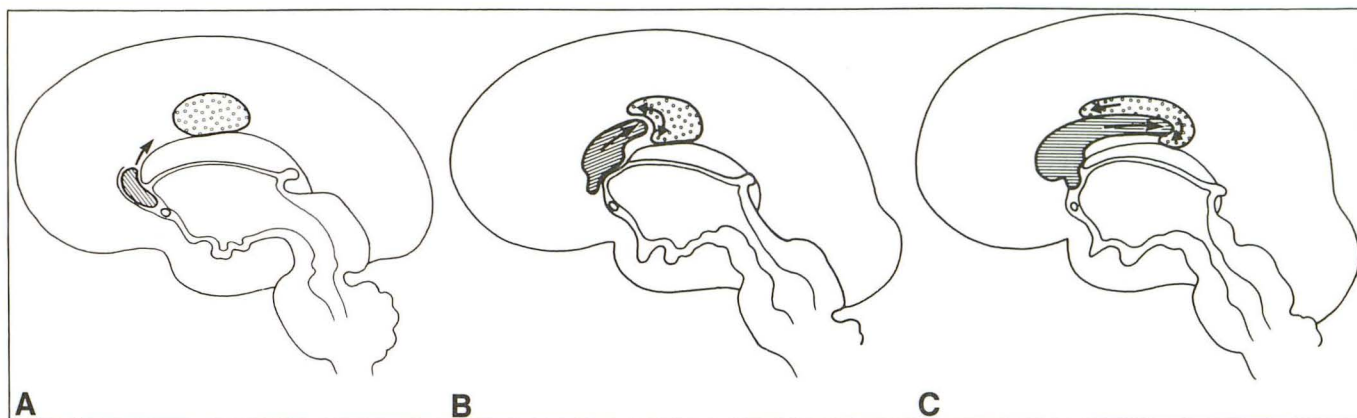


Fig. 7.—Schematics illustrating embryonic callosal collision.

A, At initiation of callosal induction at 12 weeks, a deep midline neural tube inclusion mass is seen within the path of future callosal growth (arrow).

B, With further growth, the corpus callosum eventually collides with the inclusion mass, indenting it (arrows).

C, The inclusion mass, if sufficiently pliable (i.e., lipomatous tissue), becomes invaginated by corpus callosum, resulting in individually varying degrees of obstructive callosal hypogenesis and cometlike cloaking of the callosum.

matter dysgenesis (Fig. 2C) [4, 9, 12, 13]. As callosal agenesis alone does not produce ventricular enlargement, perhaps a universal insult during fetal gestation results in a malformation spanning these three cerebral regions, terminating in a bulk hypoplasia of the hemispheric white matter [7]. In fact, polymicrogyria is usually found histopathologically in association with colpocephaly, the former of which may be radiologically occult [7, 9]. Why the occipital regions are the most severely affected is uncertain; however, the injury likely occurs later in gestation, after the frontal and anterior parietal areas have largely developed. In addition, it seems that the occipital white

matter is the last to mature within the human cerebrum, a maturation that sometimes continues into the second decade [14]. Therefore, late-occurring insults might be most pronounced in this region. Interestingly, concomitant involvement of the temporal lobes was noted in this series, has been observed by others, and should be included in the concept of colpocephaly (Fig. 2) [7, 13].

It must be stated that the dysgenetic change in the cerebral cortex is difficult to detect in MR in regard to location, extent, and precise type. This distinction must remain essentially a histopathologic one, since even gross errors in cortical lami-

nation and sulcation may only be discernible at microscopy [7, 10]. Additionally, Table 2 indicates the frequent superimposition of these gyral dysgenetic states and related colpocephaly upon the agenetic and hypogenetic conditions of the corpus callosum (Figs. 2 and 3). The classification in Table 1 also reflects this association and further indicates that, for these reasons, callosal hypoplasia can be found in combination with hypogenesis of the corpus callosum. It also follows, as previously noted, that the segmental or complete Probst bundles, which are the callosal equivalents, may be hypoplastic in cases of both hypogenesis and agenesis. Whether concomitant dysgenesis of the corpus callosum and dysgenesis of the cerebral hemispheres represent secondary anomalies due to the same fetal insult, and/or whether they exert compound bi-directional effects upon one another is uncertain. In fact, each case is a unique result of factors and influences, with the product being a complex telencephalic mosaic perhaps best termed in many cases as a corticocallosal dysgenesis [7, 9, 15].

The occurrence of midline cysts in conjunction with callosal dysgenesis has been reported in the literature and seems to relate to generalized maldevelopment, although the exact association is unknown (Fig. 3) [4, 5, 13, 16]. The observed uni- or bilateral hypoplastic cerebral peduncles in three subjects are best explained by a paucity of projection fibers emanating from the dysgenetic cerebral hemisphere(s) reflected in an underlying pyramidal tract hypoplasia (Fig. 1) [7, 15, 17].

The high association of mental retardation or developmental delay of individually variable severity in this series indicates the obvious strong dependence of function upon gross developmental structural morphology. While primary agenesis per se may not cause significant functional deficits, this anomaly is frequently allied with concomitant abnormalities within the cerebral cortex or hemispheric white matter, which will have profound effects clinically [4, 7, 11, 15, 17, 18]. The same inferences are true for interruptive hypogenesis, and also of course for cortically dependent callosal hypoplasia.

The single category of callosal dysgenesis that revealed no evidence of mental retardation was that of obstructive callosal hypogenesis. These cases of lipomatous neural tube inclusion would seem to reveal a separation of the effects of neural tube closure from those of separate insults occurring later during callosal induction, neuronal migration, and cortical lamination. The relatively benign embryonic collision of the developing corpus callosum with the inclusion mass primarily affects functions mediated by the corpus callosum itself, assuming there is no other cerebral anomaly [4, 18]. The cases of isolated obstructive hypogenesis, therefore, have features that make them largely clinically distinct in most cases from the other categories of callosal dysgenesis.

The observations in this series of patients graphically illustrate that the embryogenesis of the telencephalon dictates the final structural morphology of the corpus callosum. During normal gestation this depends on the uncomplicated closure of the neural tube, the formation and maintenance of the

inductive bed of the massa commissuralis throughout decussation, and the proper migration and lamination of the elements of the cerebral cortex. Interference with, or interruption of, any one or a combination of these parameters may result in callosal or corticocallosal dysgenesis of individually unique clinicoanatomic expression. Thus, the range of callosal or corticocallosal dysgenesis reflects its origins in an essential failure of decussation induction, interrupted decussation, and/or partial failure of commissural fiber projection secondary to various degrees of primary cortical dysgenesis.

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