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**Classification of congenital abnormalities of the  
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*AJNR Am J Neuroradiol* 1988, 9 (2) 315-326

<http://www.ajnr.org/content/9/2/315>

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# Classification of Congenital Abnormalities of the CNS

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**A classification of congenital cerebral, cerebellar, and spinal malformations is presented with a view to its practical application in neuroradiology. The classification is based on the MR appearance of the morphologic abnormalities, arranged according to the embryologic time the derangement occurred. The normal embryology of the brain is briefly reviewed, and comments are made to explain the classification. MR images illustrating each subset of abnormalities are presented.**

During the last few years, MR imaging has proved to be a diagnostic tool of major importance in children with congenital malformations of the CNS [1]. The excellent gray/white-matter differentiation and multiplanar imaging capabilities of MR allow a systematic analysis of the condition of the brain in infants and children. This is of interest for estimating prognosis and for genetic counseling.

A classification is needed to serve as a guide to the great diversity of morphologic abnormalities and to make the acquired data useful. Such a system facilitates encoding, storage, and computer processing of data. We present a practical classification of congenital cerebral, cerebellar, and spinal malformations. Our classification is based on the morphologic abnormalities shown by MR and on the time at which the derangement of neural development occurred.

A classification based on etiology is not as valuable because the various presumed causes rarely lead to a specific pattern of malformations. The abnormalities reflect the time the noxious agent interfered with neural development, rather than the nature of the noxious agent. The vulnerability of the various structures to adverse agents is greatest during the period of most active growth and development. A time can be given for all events that occur during development of the CNS. To the extent possible, we note the time at which each malformation had its onset.

## Classification

The system presented by Volpe [2] provides an excellent basis for composing a classification of congenital abnormalities of the CNS. In contrast to others and in conformity with our intention, Volpe arranged the disorders according to the time of onset of the morphologic derangement. The application of his classification in neuroradiology, however, is limited, because important conditions have been omitted, such as cerebellar malformations, congenital vascular malformations, congenital tumors, and secondarily acquired congenital abnormalities. Our classification (Table 1) is intended to be as complete as possible. Where it is incomplete, it provides a framework within which the missing malformation can be allocated.

To understand our classification, knowledge of the major events in embryologic and fetal neural development is essential. Our classification contains eight subsets, which are discussed, and the developmental events per subset are described briefly [2, 60, 61]. MR images are used to illustrate the various abnormalities. Gradient-

Received May 15, 1987; accepted after revision October 6, 1987.

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**AJNR 9:315-326, March/April 1988**  
0195-6108/88/0902-0315

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TABLE 1: Classification of Congenital Cerebral, Cerebellar, and Spinal Malformations

Disorder	Time of Onset During Gestation	Disorder	Time of Onset During Gestation
1. Dorsal induction:			
Primary neurulation/neural tube defects (3–4 weeks' gestation) [2, 3]:		3.10 Congenital vascular malformations [31]	2–3 months
1.1 Craniorachischisis totalis	3 weeks	3.11 Congenital tumors of the nervous system [31–33] <sup>a</sup>	–
1.2 Anencephaly	4 weeks	3.12 Aqueduct stenosis [2, 29, 34]	4 months
1.3 Myeloschisis	4 weeks	3.13 Colpocephaly [35, 36]	2–6 months
1.4 Encephalocele	4 weeks	3.14 Porencephaly [37–39]	3–4 months
1.5.1 Myelomeningocele	4 weeks	3.15 Multicystic encephalopathy [37, 40]	3–4 months
1.5.2 Chiari malformation	4 weeks	3.16 Hydranencephaly [41, 42]	3 months or later
1.5.3 Hydromyelia	4 weeks	4. Migration (2–5 weeks' gestation):	
Secondary neurulation/occult dysraphic states (4 weeks' gestation–postpartum period) [2, 3]:		4.1 Schizencephaly [43–45]	2 months
1.6 Myelocystocele	4 weeks	4.2 Lissencephaly [46]	3 months
1.7 Diastematomyelia/diplomyelia [4]	4–5 weeks	4.3 Pachygyria [47]	3–4 months
1.8 Meningocele/lipomeningocele	4–5 weeks	4.4 Polymicrogyria [48]	5 months
1.9 Lipoma <sup>a</sup>	–	4.5 Neuronal heterotopias [2]	5 months
1.10 Dermal sinus with or without dermoid or epidermoid cyst [5]	3–5 weeks	4.6 Hypoplasia/aplasia of the corpus callosum [2]	3–5 months
1.11 Tethered cord/tight filum terminale syndrome	4–5 weeks	5. Myelination (7 months' gestation–1 year of age):	
1.12 Anterior dysraphic disturbances	4–5 weeks	5.1 Hypomyelination [1, 2]	–
1.13 Caudal regression syndrome [6, 7]	4–7 weeks	5.2 Retarded myelination [1, 49]	–
2. Ventral induction (5–10 weeks' gestation):		6. Secondarily acquired injury of normally formed structures:	
2.1 Atelencephaly [2]	5 weeks	6.1 Encephaloclastic atelencephaly [50]	–
2.2 Holoprosencephaly [8]	5–6 weeks	6.2 Encephaloclastic hydranencephaly [51, 52]	–
2.3 Septooptic dysplasia [9, 10]	6–7 weeks	6.3 Encephaloclastic multicystic encephalopathy [53]	–
2.4 Agenesis of the septum pellucidum [11, 12]	6 weeks	6.4 Encephaloclastic schizencephaly	–
2.5 Diencephalic cyst [13, 14]	6 weeks	6.5 Encephaloclastic porencephaly [31]	–
2.6 Cerebral hemihypoplasia/aplasia [15]	6 weeks	6.6 Aqueduct stenosis [29]	–
2.7 Lobar hypoplasia/aplasia [15, 16]	6 weeks	6.7 Hydrocephalus	–
2.8 Hypoplasia/aplasia of the cerebellar hemispheres [17]	6–8 weeks	6.8 Damage of the corpus callosum	–
2.9 Hypoplasia/aplasia of the vermis cerebelli including Joubert syndrome [18–20]	6–10 weeks	6.9 Defect of the septum pellucidum [11]	–
2.10 Dandy-Walker syndrome and Dandy-Walker variant [21–23]	7–10 weeks	6.10 Sclerotic polymicrogyria [2, 54]	–
2.11 Craniostyostosis [24, 25]	6–8 weeks	6.11 Cerebral atrophy	–
3. Neuronal proliferation, differentiation, and histogenesis (2–5 months' gestation):		6.12 Cerebellar atrophy	–
3.1 Micrencephaly [2, 26]	2–4 months	6.13 Demyelination	–
3.2 Megalencephaly [2, 27]	2–4 months or later	6.14 Congenital cerebral calcifications [55]	–
3.3 Unilateral megalencephaly [28]	2–4 months or later	6.15 Hemorrhage in the plexus	–
3.4 Von Recklinghausen disease [12, 29, 30]	5 weeks–6 months	6.16 Subependymal hemorrhage	–
3.5 Bourneville disease [12, 29, 30]	"	6.17 Other parenchymatous hemorrhage	–
3.6 Sturge-Weber disease [12, 29, 30]	"	6.18 Subdural hematoma	–
3.7 Von Hippel-Lindau disease [12, 29, 30]	"	6.19 Subdural effusion [56]	–
3.8 Ataxia telangiectasia [12, 29, 30]	"	6.20 Periventricular leukomalacia	–
3.9 Other neurocutaneous syndromes [30]	"	6.21 Congenital infarction	–
		7. Degenerative diseases of the developing nervous system [2]:	
		7.1 Primarily affecting gray matter	–
		7.2 Primarily affecting white matter/dysmyelination	–
		8. Unclassified:	
		8.1 Arachnoid cysts [31, 57–59]	–
		8.2 Other	–

<sup>a</sup> The time of onset for this disorder cannot be pinpointed within the range given.

echo, spin-echo (SE), and inversion-recovery (IR) images were obtained with varying repetition and echo times. In the figure legends, SE 350/30 reflects a repetition time of 350 msec and echo time of 30 msec; IR 2400/600 reflects a repetition time of 2400 msec and an inversion time of 600 msec.

#### Subset 1: Disorders of Dorsal Induction

The development of the human nervous system commences with the development of the notochordal process, which extends from the primitive knot to the cranial end of

the embryo. The notochordal process induces the neural plate, a thickened area of embryonic ectoderm. The neural plate forms the neural tube, which eventually gives rise to the spinal cord and brain. These inductive events are referred to as *dorsal induction*. In these processes distinction is made between primary and secondary neurulation. *Primary neurulation* refers to the formation of the neural tube from approximately the L1 or L2 level, which corresponds to the caudal end of the notochord upward to the cranial end of the embryo. These events occur during the third and fourth weeks of gestation. *Secondary neurulation* refers to the formation of the caudal neural tube below the caudal end of the notochord by canalization and retrogressive differentiation. The lower lumbar, sacral, and coccygeal segments are thus formed. This canalization starts at 4 weeks and continues until 7 weeks of gestation. The retrogressive differentiation lasts from the seventh week until some time after birth.

Disturbances of dorsal induction result in the disorders listed in subset 1 of our classification. Several features of these disorders are noteworthy:

1. Myelomeningoceles, Chiari malformations, and hydromyelia are so often associated with each other that they are considered to be one clinical syndrome. The syndrome may be manifest with complete or incomplete expression [62–65].
2. The distinction is unclear between derangements of primary and secondary neurulation. The Chiari malformation is sometimes found with diastematomyelia [66, 67], and diastematomyelia is sometimes found in the cervical and thoracic regions [4, 68]. Disorders of primary and secondary neurulation have been found to occur in one family in a higher than average percentage [69].
3. Less common, though related, lesions include anterior dysraphic disturbances, such as neurenteric cyst and anterior meningocele.

One of the failures of dorsal induction is illustrated in Figure 1.

#### Subset 2: Disorders of Ventral Induction

Ventral induction refers to the inductive events occurring ventrally in the rostral end of the embryo, resulting in the formation of the face and brain. The most important events involve formation of the prosencephalon, mesencephalon, and rhombencephalon. The prosencephalon divides into telencephalon and diencephalon, the rhombencephalon into metencephalon and myelencephalon. These give rise to the various cerebral and cerebellar structures. The telencephalon divides into two parts, resulting in two hemispheres and two lateral ventricles. The hemispheres roll over, and as a consequence the temporal lobes and lateral fissures are formed. The two hemispheres are joined together by the corpus callosum. Major events occur between the fifth and tenth weeks of gestation. The cerebellum develops from the dorsal part of the metencephalon. At the beginning of the second month of embryonic life, rapid growth takes place at both sides of the midline. At the end of the second month, the cerebellar rudiments of the two sides merge in the midline. Once fusion is complete, the primordial cerebellum grows downward and backward.

The derangements of ventral induction are listed in subset 2 of our classification. A few items are noteworthy:

1. In contrast to Volpe [2], we include the congenital malformations of the posterior fossa in this section, since we are convinced that the ventral inductive events comprise not only the formation of the prosencephalic structures, but also of the mesencephalic and rhombencephalic structures.
2. The cerebellar vermis develops in a rostrocaudal direction, as is the case with the corpus callosum. In developmental

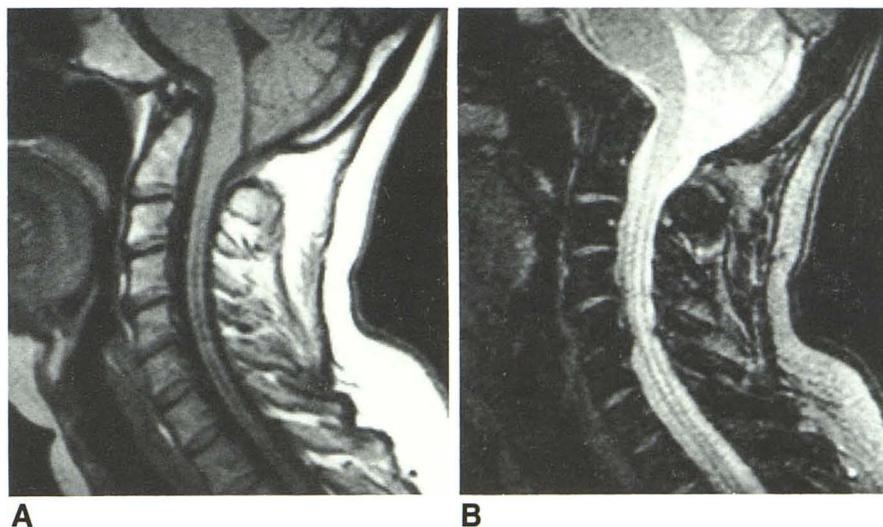


Fig. 1.—Classification: Chiari malformation (1.5.2) and hydromyelia (1.5.3). SE 350/30 image with six excitations (A) and gradient-echo 150/30 image with eight excitations and 25° flip angle (B) show forme fruste of Chiari II malformation, with medullary kinking and descent of cerebellar tonsils and vermis combined with bony abnormalities of craniovertebral junction, short clivus, high position of dens, assimilation of posterior arch of atlas, and small hydromyelic cavity beginning at C2 level and continuing downward. Gradient-echo image (B) shows hydromyelia to better advantage.

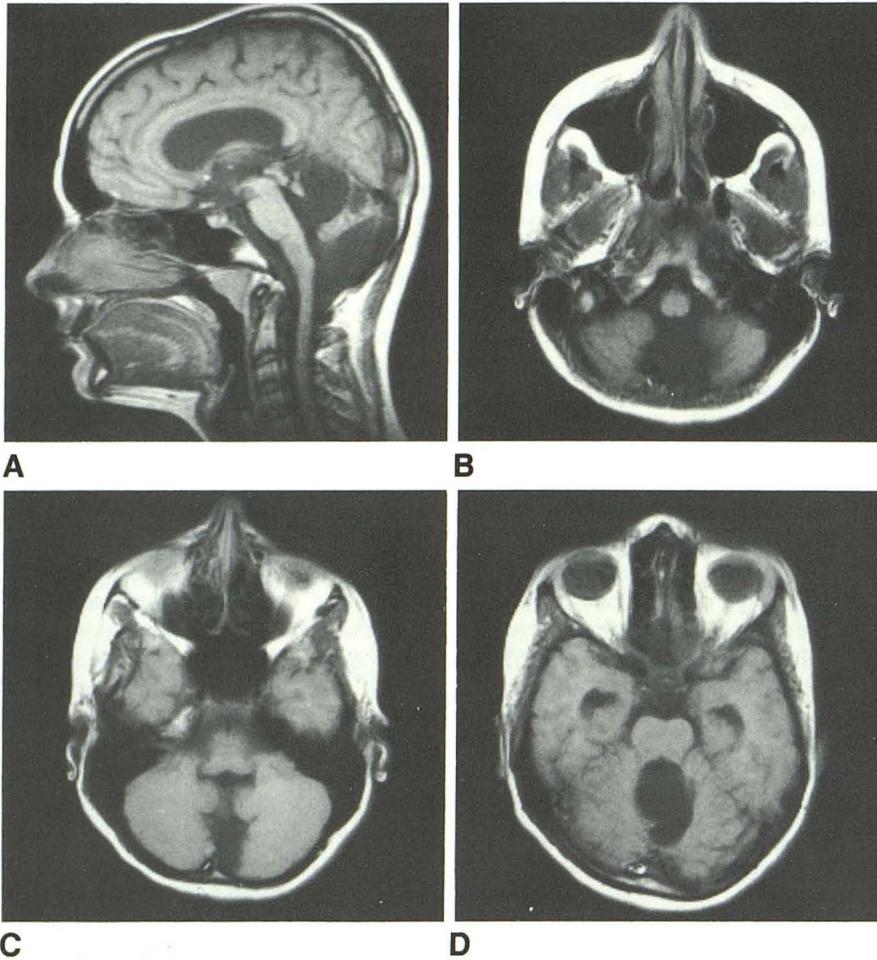


Fig. 2.—Classification: hypoplasia/aplasia of vermis cerebelli including Joubert syndrome (2.9). SE 350/35 images with six excitations and  $192 \times 256$  matrix.

A, Midsagittal plane shows complete absence of cerebellar vermis.

B–D, Transverse slices confirm agenesis of cerebellar vermis with normal development of cerebellar hemispheres. Well-developed cerebellar hemispheres indicate that this case should not be classified as Dandy-Walker cyst or as Dandy-Walker variant. True vermis aplasia, as seen here, is a very rare disorder.

disorders, either the entire cerebellar vermis or the caudal part is absent or hypoplastic. When the rostral part is undersized or absent, the damage must have been acquired secondarily.

3. Craniosynostosis appears to result from an early embryonic disturbance in the formation of the cranial base of the skull and as such can be included in the disorders of ventral induction [24, 25].

Figure 2 illustrates a failure of ventral induction.

#### *Subset 3: Disorders of Neuronal Proliferation, Differentiation, and Histogenesis*

Once the essential external form of the brain has been established, complex processes of neuronal proliferation, differentiation, migration, and organization follow. Major events initially occur between 2 and 5 months of gestation, although after this time processes continue into the postnatal period.

Disorders of neuronal proliferation, differentiation, and histogenesis are considered as one subset, as in most diseases two or three of these processes are involved. Although migrational processes occur simultaneously, in a number of conditions pathology is predominantly a derangement of mi-

gration; therefore, these malformations are classified as a separate subset. Features worth highlighting are listed:

1. Various types of tumors have been found to be present in the CNS at birth [32]. Some tumors are related to the remains of embryonic neural cells, for example, craniopharyngiomas, chordomas, medulloblastomas, and hamartomas. Other tumors originate from residual embryonic cells normally present in the cranial cavity, but alien to nervous tissue; for example, teratomas, germinomas, dermoids, and epidermoids. Also tumors that are not of developmental origin have been found at birth; for example, ependymomas, astrocytomas, and oligodendrogliomas. The time of onset of the developmental derangement will differ for the various tumors depending on type. The causes vary from genetic influences to teratogens and oncogens.

2. The origin of congenital vascular malformations, such as arteriovenous malformations, congenital aneurysms, and angiomas, dates back to early developmental stages, but the precise time of onset is controversial and possibly differs for the various types of vascular abnormalities.

3. The precise origin and onset of the neurocutaneous disorders is speculative and has been addressed [12, 29, 30].

4. We are not sure whether aqueduct stenosis, colpocephaly, porencephaly, multicystic encephalopathy, and hydranencephaly are disorders of neuronal proliferation and histogenesis, although this viewpoint is certainly tenable. We placed them in this subset because of their times of onset.

Figures 3 and 4 illustrate disorders of neuronal proliferation, differentiation, and histogenesis.

#### Subset 4: Disorders of Migration

Neurons are generated in the ventricular and subventricular layers of the brain. Migration of the nerve cells from their site of origin to the superficial cortex and deep nuclei of the brain occurs predominantly in the third, fourth, and fifth months of gestation. Likewise, in this period the cerebellar nuclei and cortex are formed. Disorders of neuronal migration result in an abnormal gyral pattern, ranging from very few thick gyri or

no gyri to an excess of very small gyri. In addition, in migrational disorders the corpus callosum is often hypoplastic or absent, as the development of this major interhemispheric commissure is associated with cerebral migrational events. Figures 5–7 illustrate, among other anomalies, disturbances of migration.

#### Subset 5: Disorders of Myelination

Normal myelination [70–72] in the brain starts in the third trimester of pregnancy and continues into adult life. At the beginning of the third trimester, myelin appears in the brainstem and in the central parts of the cerebellum. Subsequently, the myelin is formed in the thalamus and the posterior limb of the internal capsule. At birth or shortly thereafter, myelin has spread to the cerebellar hemispheres, the optic radiation, and the centrum semiovale. During the first year of life myelin

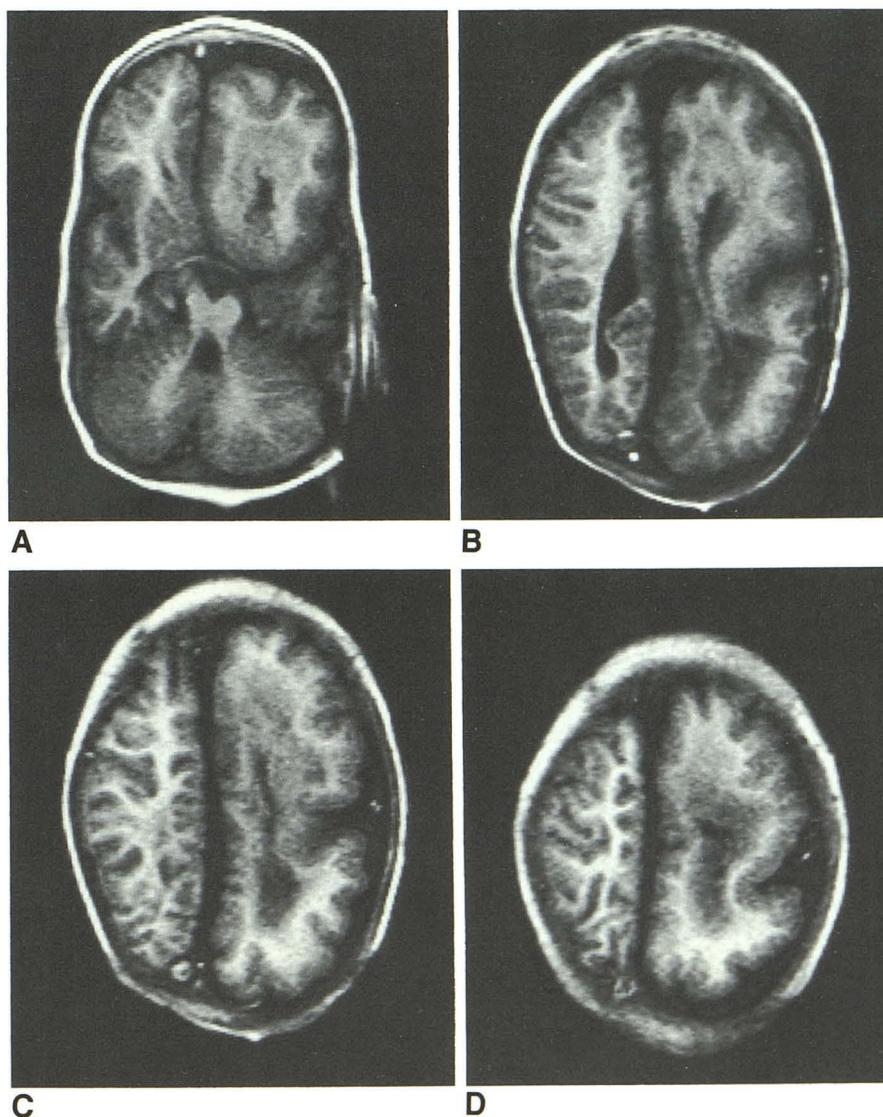


Fig. 3.—Classification: unilateral megalencephaly (3.3). IR 2400/600 images with two excitations and  $128 \times 256$  matrix show difference between two hemispheres. Right hemisphere has developed relatively normally; left one is larger and shows disfiguration of ventricular system, an irregular, thick layer of gray matter, and heterotopic gray matter. Hemimegalencephaly is an extremely rare disorder, in contrast to megalencephaly, which is found more often in diseases such as tuberous sclerosis and neurofibromatosis. Hemimegalencephaly represents a spectrum of disorders. An increased size of otherwise normal-appearing cortical neurons and mild astrocytosis is found at one end of the spectrum, while at the other end, scattered giant neurons accompanied by multitudes of hyperplastic astrocytes are found with loss of normal cortical architecture and varying degrees of migrational derangements [28]. Since the increase in the number of cells, especially astrocytes, is the most constant and conspicuous finding in all variants of hemimegalencephaly, it is classified as a disorder of proliferation.

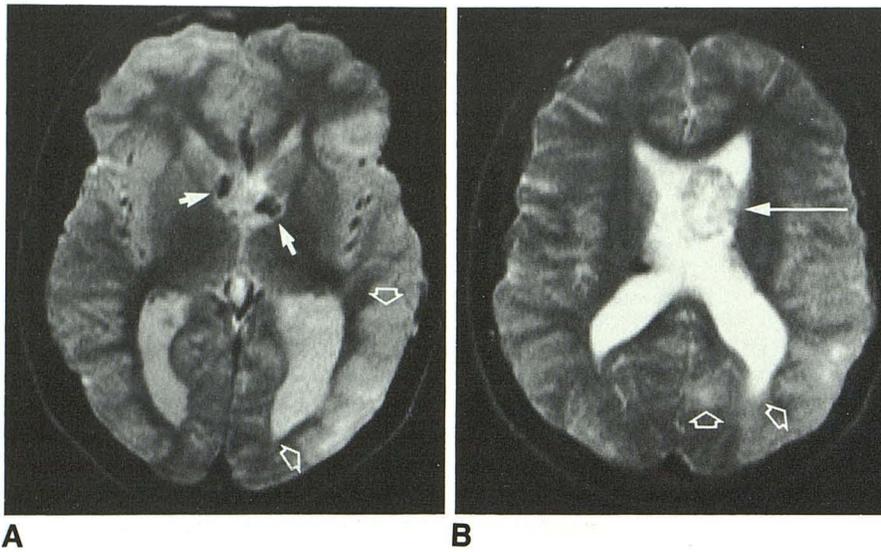


Fig. 4.—Classification: Bourneville disease (3.5). SE 3000/60 (A) and SE 3000/90 (B) images with two excitations and 128 × 256 matrix show intraventricular giant cell astrocytoma (long arrow) with subependymal calcified tubers (short solid arrows) and regions of focal gliosis (open arrows).

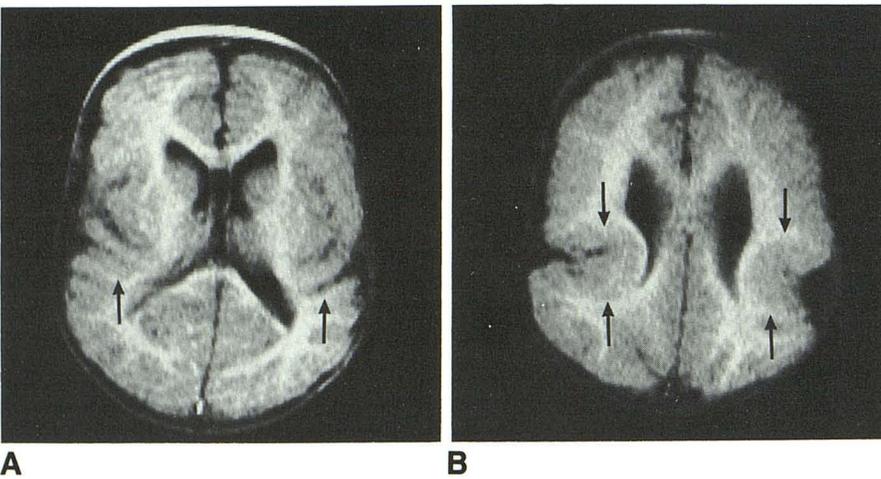


Fig. 5.—Classification: schizencephaly (4.1). SE 2000/35 images with two excitations and 128 × 256 matrix help differentiate between developmental schizencephaly and encephaloclastic schizencephaly. In developmental schizencephaly, bilateral symmetrical clefts and thick cortical layer result from migrational disorder (arrows). There are two types of developmental schizencephaly, one with open and one with fused lips. In schizencephaly with open lips the borders of the cleft are more or less widely separated. With fused lips, as shown here, the borders of the cleft are in contact, and on both sides there is a thick layer of ectopic gray matter. In developmental schizencephaly the clefts are nearly always bilateral and symmetric, located parallel to the great cerebral fissures [43–45].

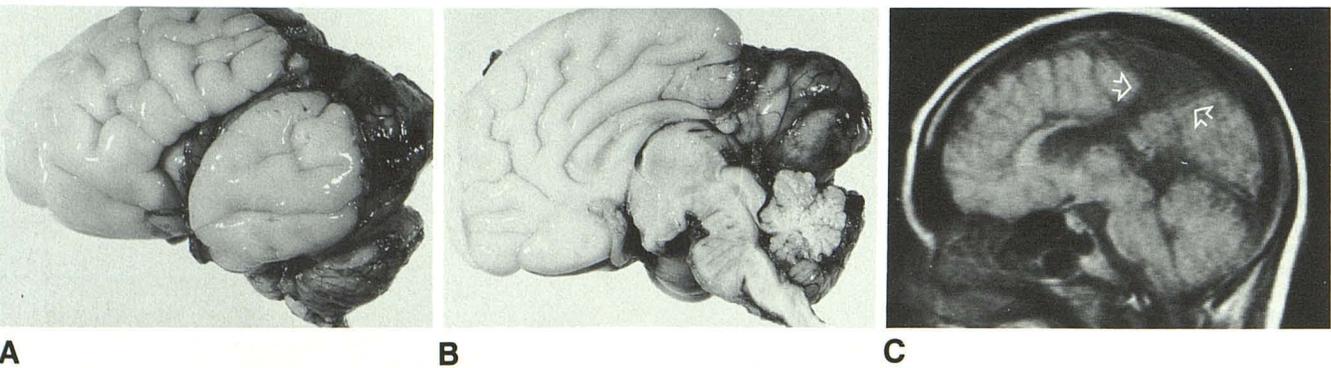
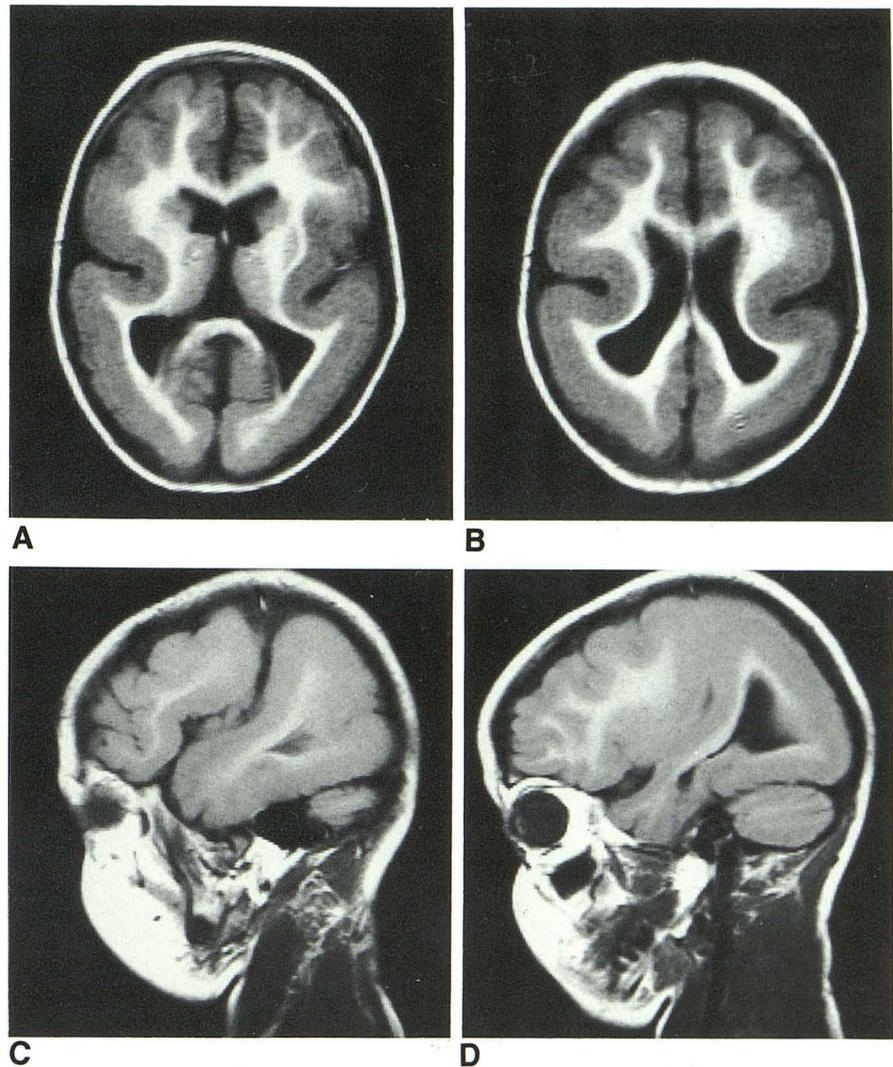


Fig. 6.—Classification: encephaloclastic schizencephaly (6.4). Cleft is often, though not always, unilateral. Signs of migrational disorder are absent. Lateral (A) and medial (B) views of anatomic specimen with classic variety of schizencephaly concur with MR image (C). Arrows indicate cleft.

In encephaloclastic schizencephaly, location of cleft depends on point of impact of noxious agent, which is why cleft is often unilateral. As encephaloclastic schizencephaly is an acquired disease after a primarily normal neural development, signs of a migrational disorder are absent [43].

Fig. 7.—Classification: pachygyria (4.3). IR 2000/500 images with two excitations and  $128 \times 256$  matrix (A and B) and SE 300/30 images with six excitations and  $192 \times 256$  matrix (C and D) show fetal stage of sylvian fissure with its vertical orientation and triangular form. This triangular form results from arrest of opercularization. Lack of gyri and thick layer of cortex are from neuronal migrational derangement in pachygyria.



spreads throughout the entire brain, though progressively finer branching of the subcortical white matter continues until early adult life.

The vulnerability of myelin to adverse external factors is increased in the period of active myelination. Stress factors will vary in their effect on myelin, producing either retardation of myelination or permanent deficit depending on its timing in relation to the process of myelination, its nature, and its severity [49].

Delayed or deficient myelination is a consequence of various abnormal conditions, including malnutrition, inborn errors of metabolism, congenital infections, and hydrocephalus [1, 2]. The diagnosis of retarded myelination vs hypomyelination can be made only with the help of repeated MR imaging. MR findings suggest that decreased myelination is often from a delay rather than a deficiency of myelination [1]. MR provides the possibility of assessing the progress of myelination in living infants, as shown in Figure 8.

#### *Subset 6: Secondarily Acquired Injury of Normally Formed Structures*

Subset 6 comprises the disorders in which structures that initially developed normally and were normally endowed become damaged secondarily. In these disorders, the noxious agent was present at a later stage of gestation, after completion of normal development [73].

Sometimes the distinction between a developmental and encephaloclastic disorder can be made on morphologic grounds, as is shown in schizencephaly. Usually, however, the distinction has to be made on the basis of clinical history and other circumstantial evidence. With the help of these data it is often possible to allocate the right classification number. A few items in this subset are noteworthy:

1. Some morphologic abnormalities arise either early in gestation as a developmental derangement, or later on as

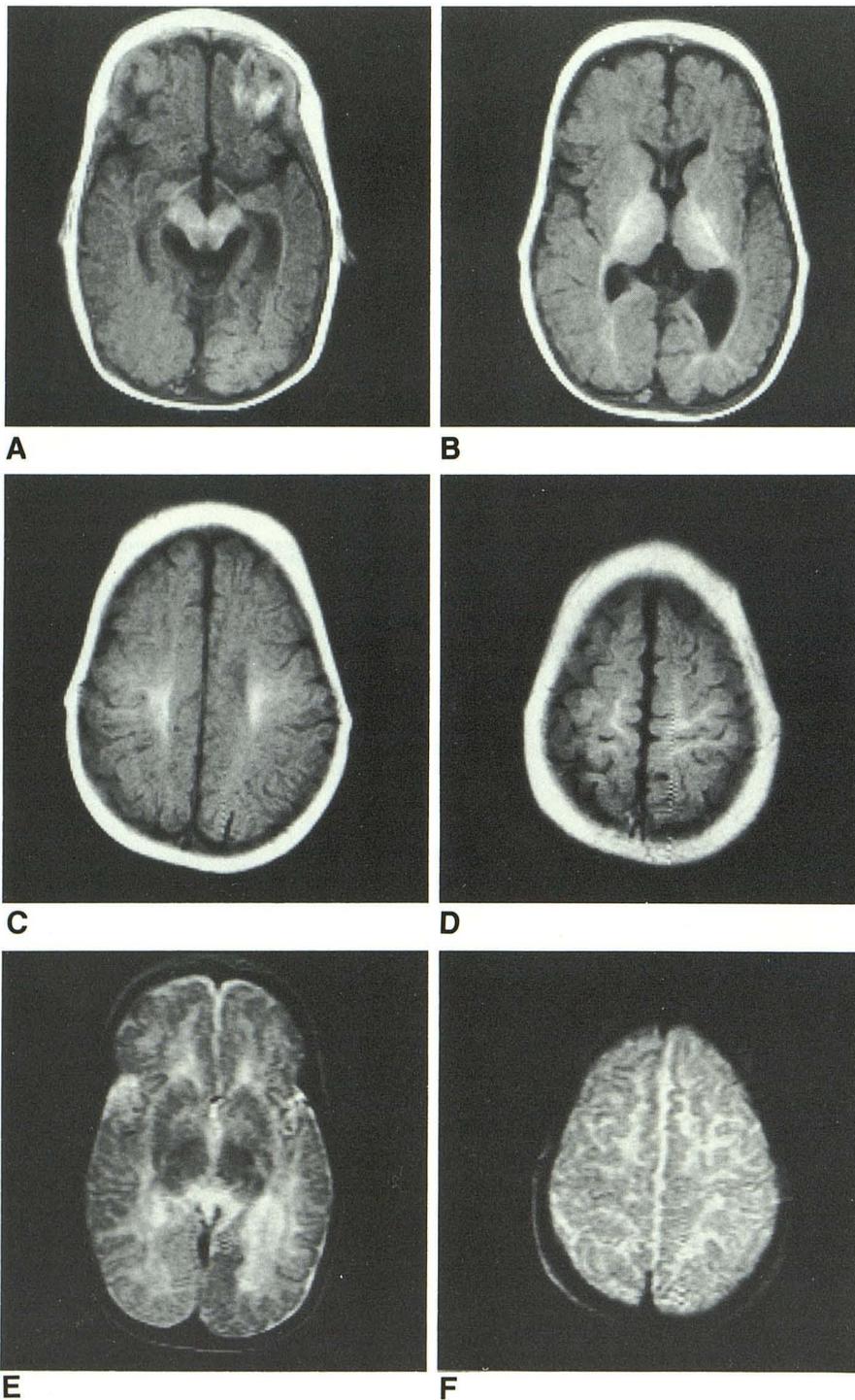


Fig. 8.—Classification: retarded myelination (5.2.).

A–D, 8-month-old child. IR 2400/600 images with two excitations and  $192 \times 256$  matrix show delayed myelination. Only areas in which process of myelination has started are bright: cerebellum, brainstem, white matter of thalamus, posterior limb of internal capsule, and band of cortex of postcentral gyrus.

E and F, SE 3000/120 images with two excitations and  $128 \times 256$  matrix form counterpart of IR images by showing inversion of gray- and white-matter intensity compared with mature myelination in areas not myelinated. Myelinated areas are dark.

encephaloclastic disturbances (6.1–6.5 and 6.10) or otherwise as secondarily acquired lesions (6.6).

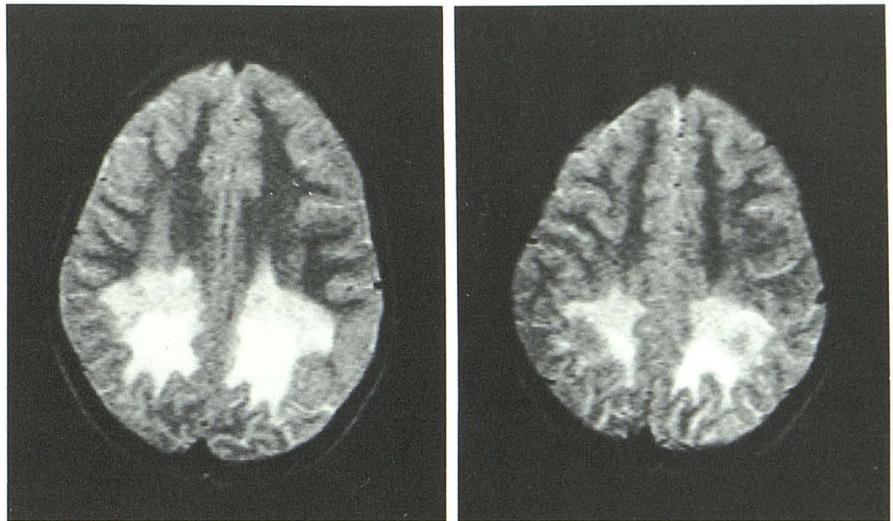
2. Usually the term *schizencephaly* is used for developmental derangements, while all encephaloclastic cavities and clefts in the brain are called *porencephalies* [45, 73]. However, we decided to distinguish between schizencephaly and porencephaly on morphologic grounds. We reserve schizence-

phaly for clefts in the brain, with or without hydrocephalus, with or without fused lips, either unilateral or bilateral, and either developmental or encephaloclastic. Porencephaly is reserved for a cavity within the cerebral hemisphere that does not interconnect the lateral ventricle and the subarachnoid space.

3. In our opinion, hydrocephalus can best be considered a

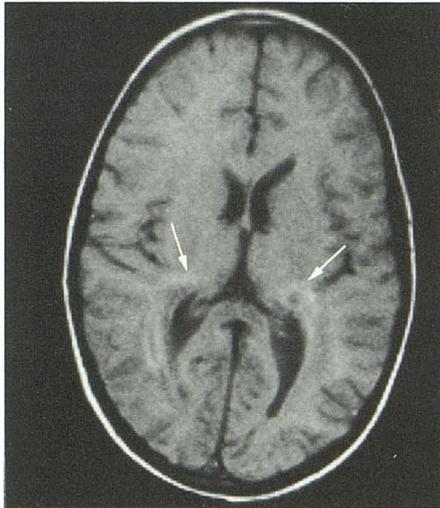
Fig. 9.—Classification: degenerative disease of developing nervous system primarily affecting white matter/dysmyelination (7.2).

Adrenoleukodystrophy. *A* and *B*, SE 3000/60 images with two excitations and  $128 \times 256$  matrix show involvement of occipitoparietal parts of white matter. *C–H*, T1-weighted images: SE 350/35, four excitations,  $192 \times 256$  matrix (*C–E*) and IR 1400/400, two excitations,  $128 \times 256$  matrix (*F–H*). Affected areas are hypointense, and active borders of disease are more hyperintense after injection of gadolinium-DTPA (arrows).



A

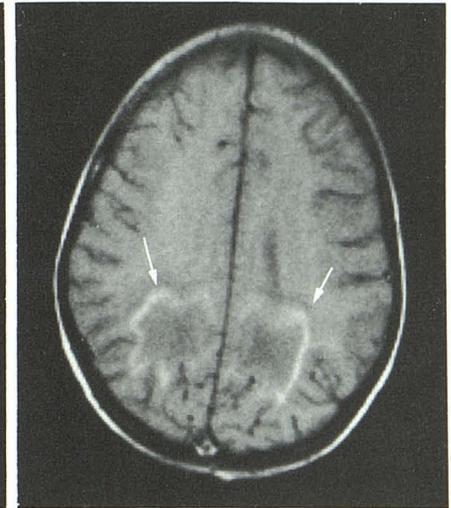
B



C



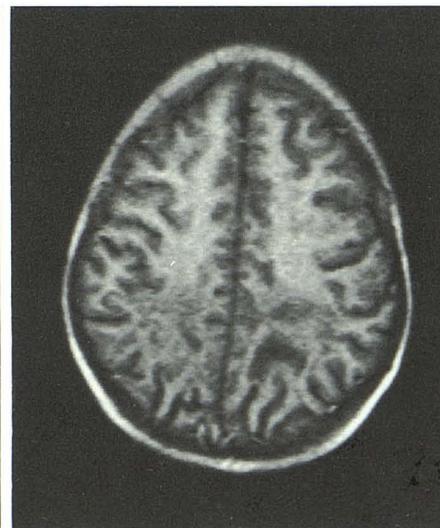
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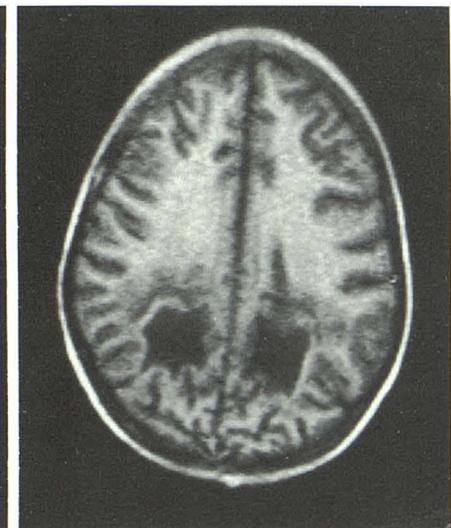
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G



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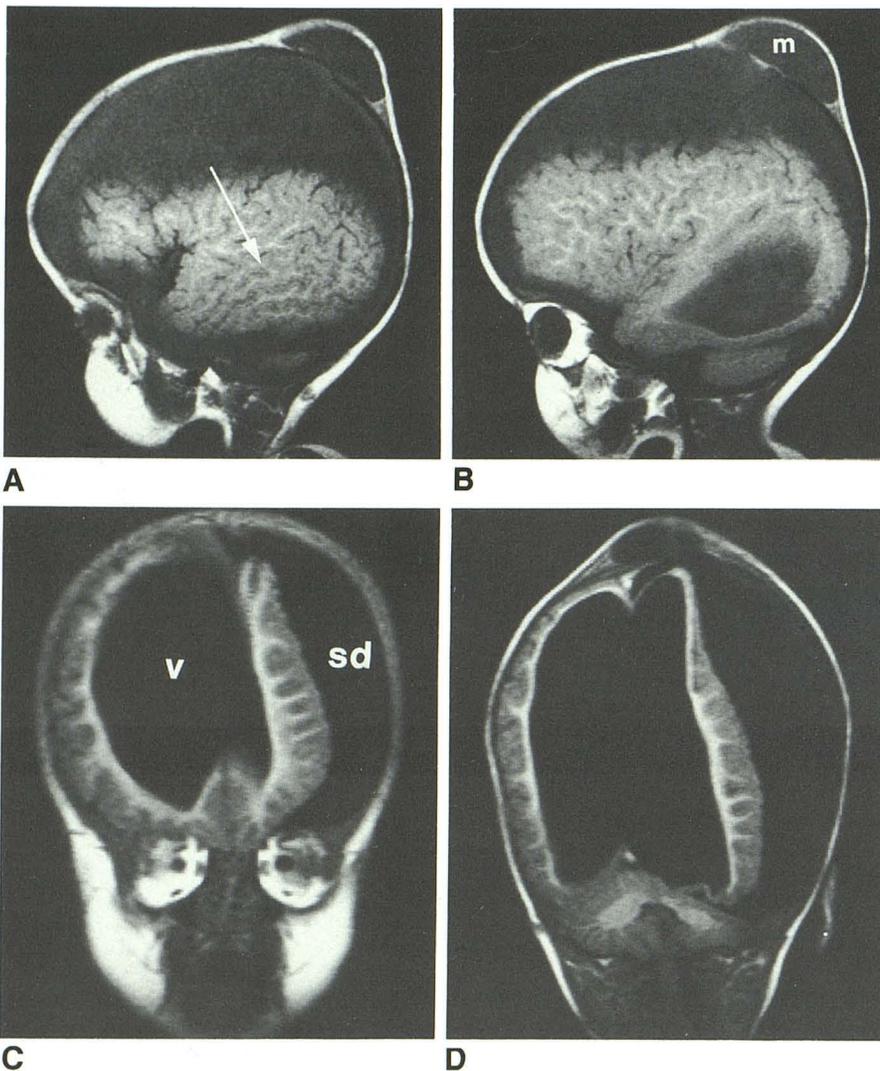


Fig. 10.—Case difficult to classify because it contains elements of more than one subset.

A and B, Sagittal SE 350/35 images with four excitations and  $192 \times 256$  matrix show enlarged skull with extensive subdural fluid collection and meningocele (m). Pattern of sulci and gyri in temporal lobe is best described as polymicrogyria (arrow).

C and D, Coronal IR 2400/600 images with two excitations and  $128 \times 256$  matrix reveal displaced monoventricle (v) and subdural fluid collection on left side (sd), apparently connected on top with meningocele (D).

Rather than force this case into subsets 4.4 (polymicrogyria) or 6.10 (sclerotic polymicrogyria) or into 2.2 (holoprosencephaly) or 6.19 (subdural hematoma), we listed it under 8.2 (other classified disorders), with the possibility of re-viewing it when more data become available.

secondarily acquired disease, because either a developmental malformation or an acquired disease underlies the hydrocephalus.

Figure 6 shows an example of encephaloclastic schizencephaly.

#### *Subset 7: Degenerative Diseases of the Developing Nervous System*

Certain degenerative diseases of the CNS may be manifest in the neonatal period, and because morphologic abnormalities may be shown by neuroradiologic investigations, especially by MR imaging, they are discussed in this context. Besides, even if these disorders become manifest some years later, they have to be regarded as developmental aberrations, which, in principle, were already present prenatally. Specific points are listed:

1. Degenerative diseases primarily affecting gray matter include the gangliosidoses, Niemann-Pick disease, and Alper disease.

2. Degenerative diseases primarily affecting white matter, the dysmyelinations, include the various leukodystrophies.

Figure 9 illustrates dysmyelination.

#### *Subset 8: Unclassified Disorders*

Subset 8 comprises those disorders in which morphologic malformations are seen that cannot be classified specifically. An example may help clarify malformations of this nature (Fig. 10).

Subset 8 includes arachnoid cysts because their pathogenesis is controversial, and it is unclear whether they are developmental or acquired. It is postulated that an inflammatory process or (minor) traumatic injury would result in adhesion

of the arachnoid membrane to the surrounding meningeal membranes. Part of the arachnoid cavity would then be excluded and behave like a cystic cavity. An alternative hypothesis suggests that an aberration in the flow of CSF during early stages of embryologic development of the subarachnoid space may lead to the formation of a blind pocket within the arachnoid membrane, a potential arachnoid cyst. Both hypotheses may be true; a distinction may be made between primary cysts from a developmental anomaly in the histogenesis of the leptomeninges and secondarily acquired cysts from adhesions [31, 57–59].

The histogenesis of the meningeal membranes is complex and takes a long time during gestation. It is not possible to pinpoint the precise time of onset of the developmental derangement resulting in an arachnoid cyst, but 3–6 months seems to be a reasonable estimation [31].

### Conclusions

Our classification can be used as a guide to the many varieties of congenital abnormalities of the CNS. It helps to structure the interpretation of complex morphologic changes and to locate them along a temporal axis. By shedding light on when the aberration commences, this classification may help to disclose the cause.

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