



## **Classification of congenital abnormalities of the CNS.**

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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-

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

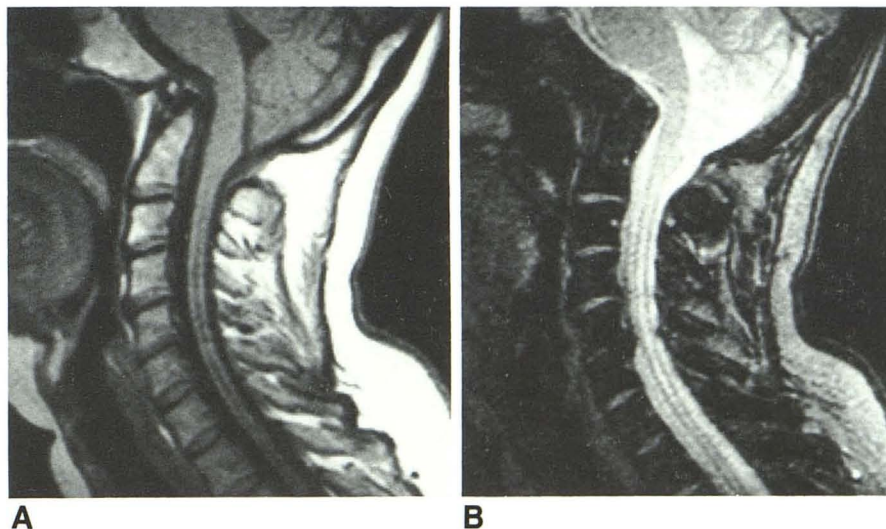
#### Subset 2: Disorders of Ventral Induction

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 cerebral 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.





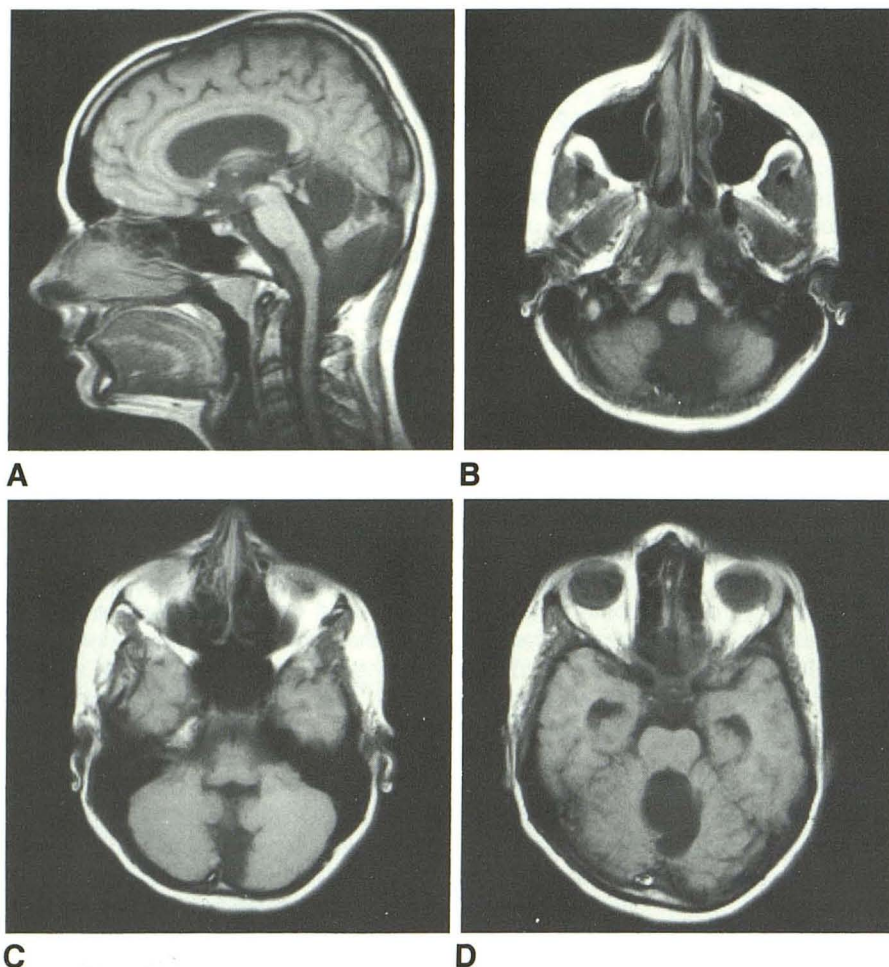


Fig. 2.—Classification: hypoplasia/aplasia of vermis cerebelli including Joubert syndrome (2.9). SE 350/35 images with six excitations and  $192 \times 256$  matrix.

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

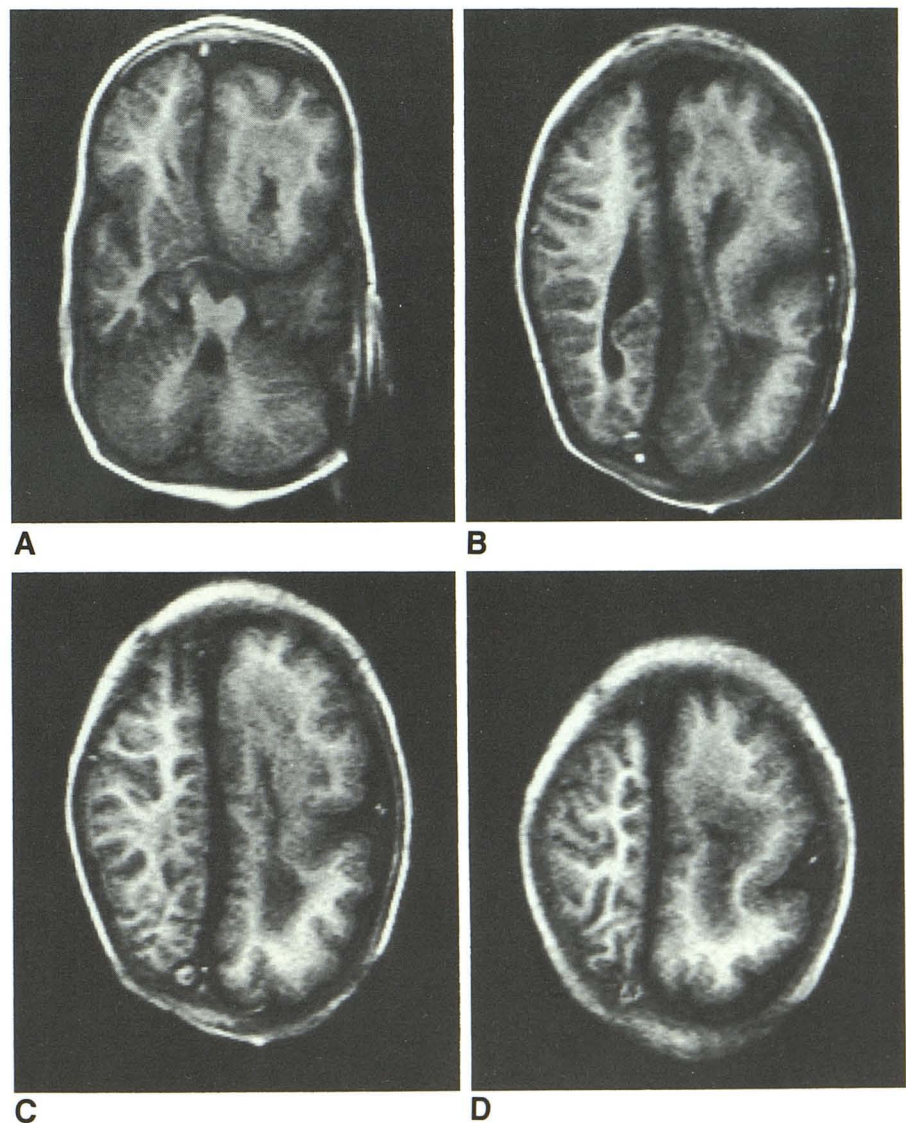


Fig. 3.—Classification: unilateral megalencephaly (3.3). IR 2400/600 images with two excitations and  $128 \times 256$  matrix show difference between two hemispheres. Right hemisphere has developed relatively normally; left one is larger and shows disfiguration of ventricular system, an irregular, thick 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.



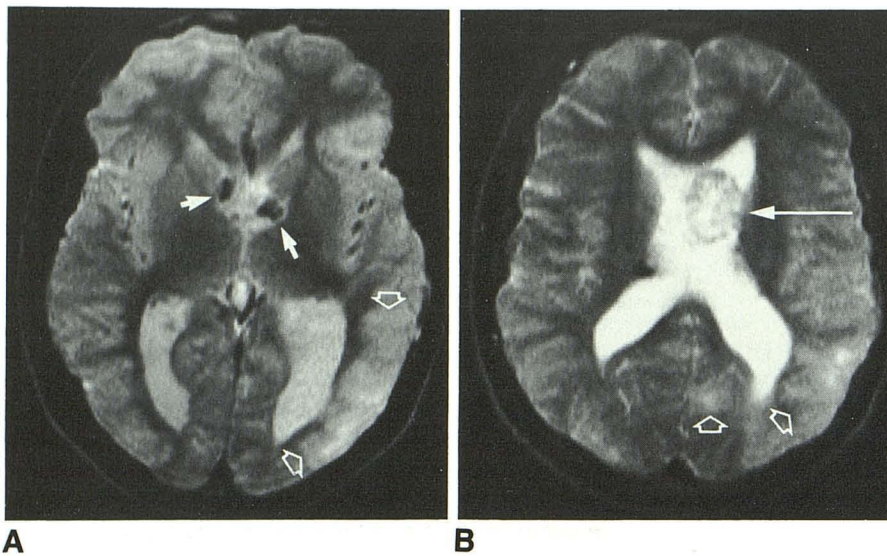


Fig. 4.—Classification: Bourneville disease (3.5). SE 3000/60 (A) and SE 3000/90 (B) images with two excitations and 128 × 256 matrix show intraventricular giant cell astrocytoma (long arrow) with subependymal calcified tubers (short solid arrows) and regions of focal gliosis (open arrows).

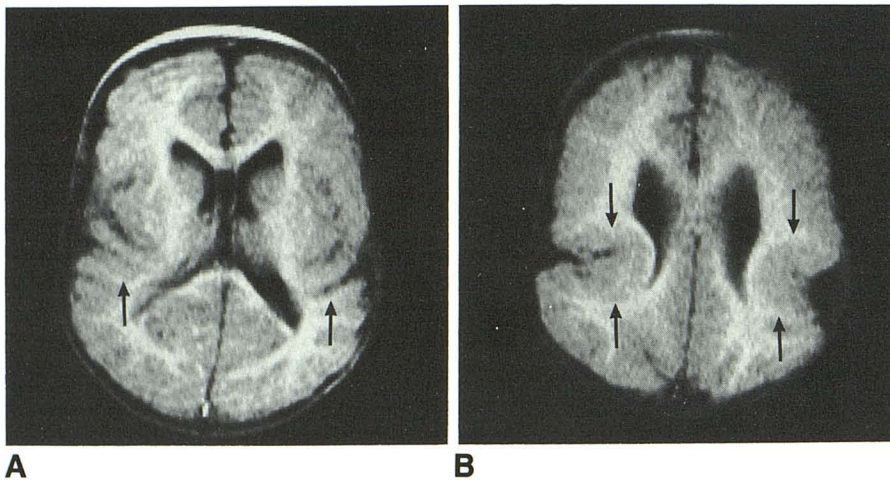


Fig. 5.—Classification: schizencephaly (4.1). SE 2000/35 images with two excitations and 128 × 256 matrix help differentiate between developmental schizencephaly and encephaloclastic schizencephaly. In developmental schizencephaly, bilateral symmetrical clefts and thick cortical layer result from migrational 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].

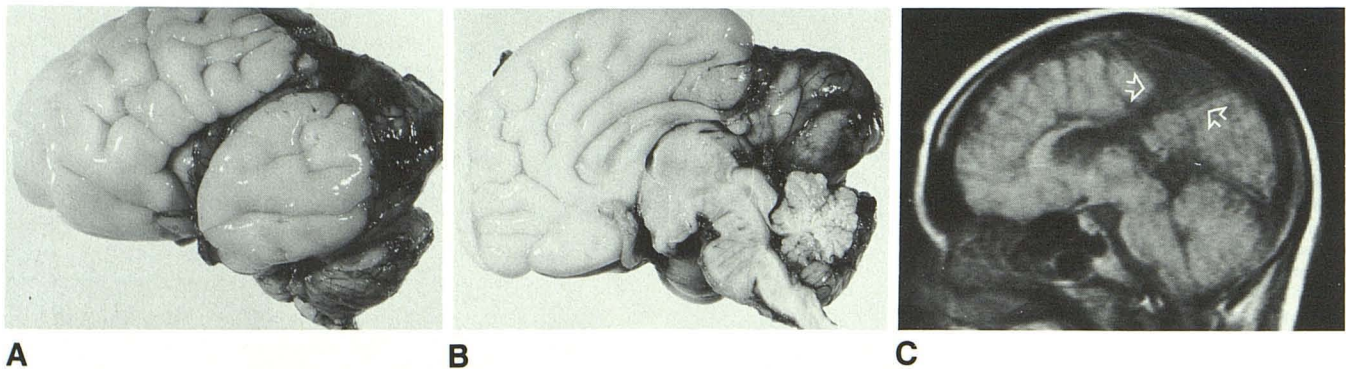
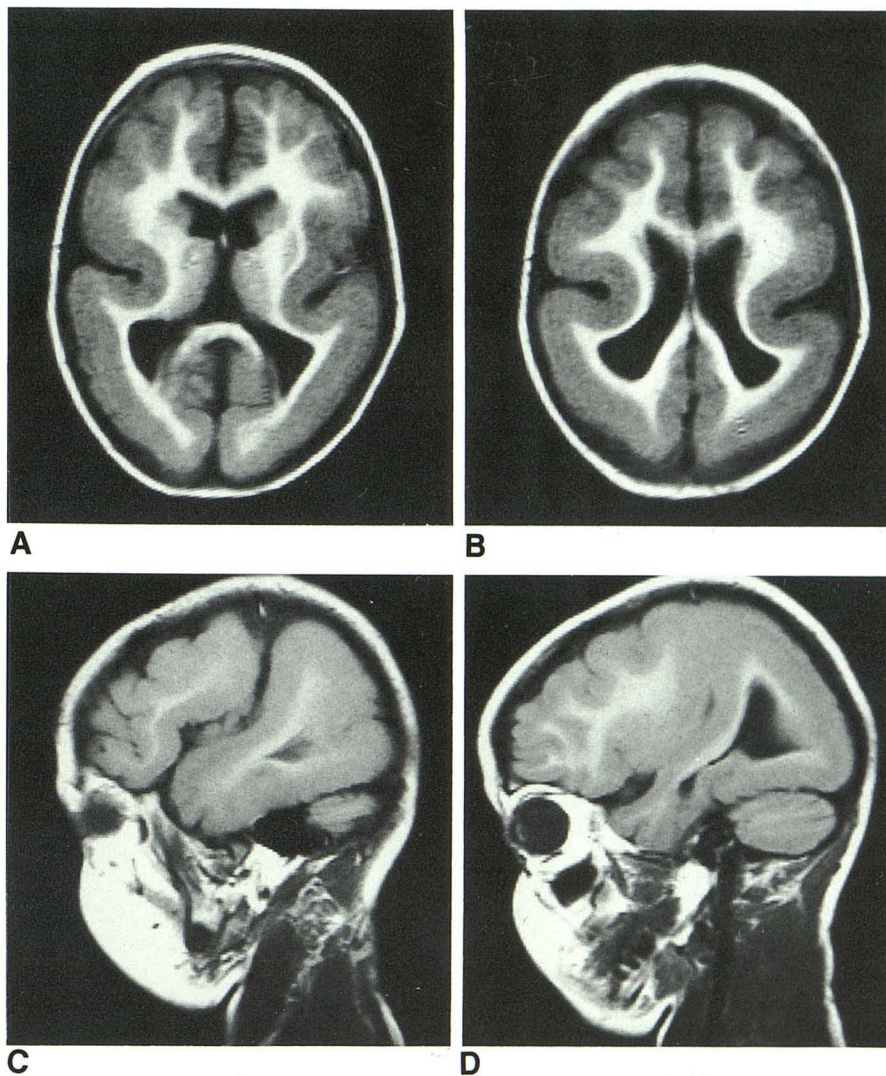


Fig. 6.—Classification: encephaloclastic schizencephaly (6.4). Cleft is often, though not always, unilateral. Signs of migrational disorder are absent. Lateral (A) and medial (B) views of anatomic specimen with classic variety of schizencephaly concur with MR image (C). Arrows indicate cleft.

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: Secondly 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



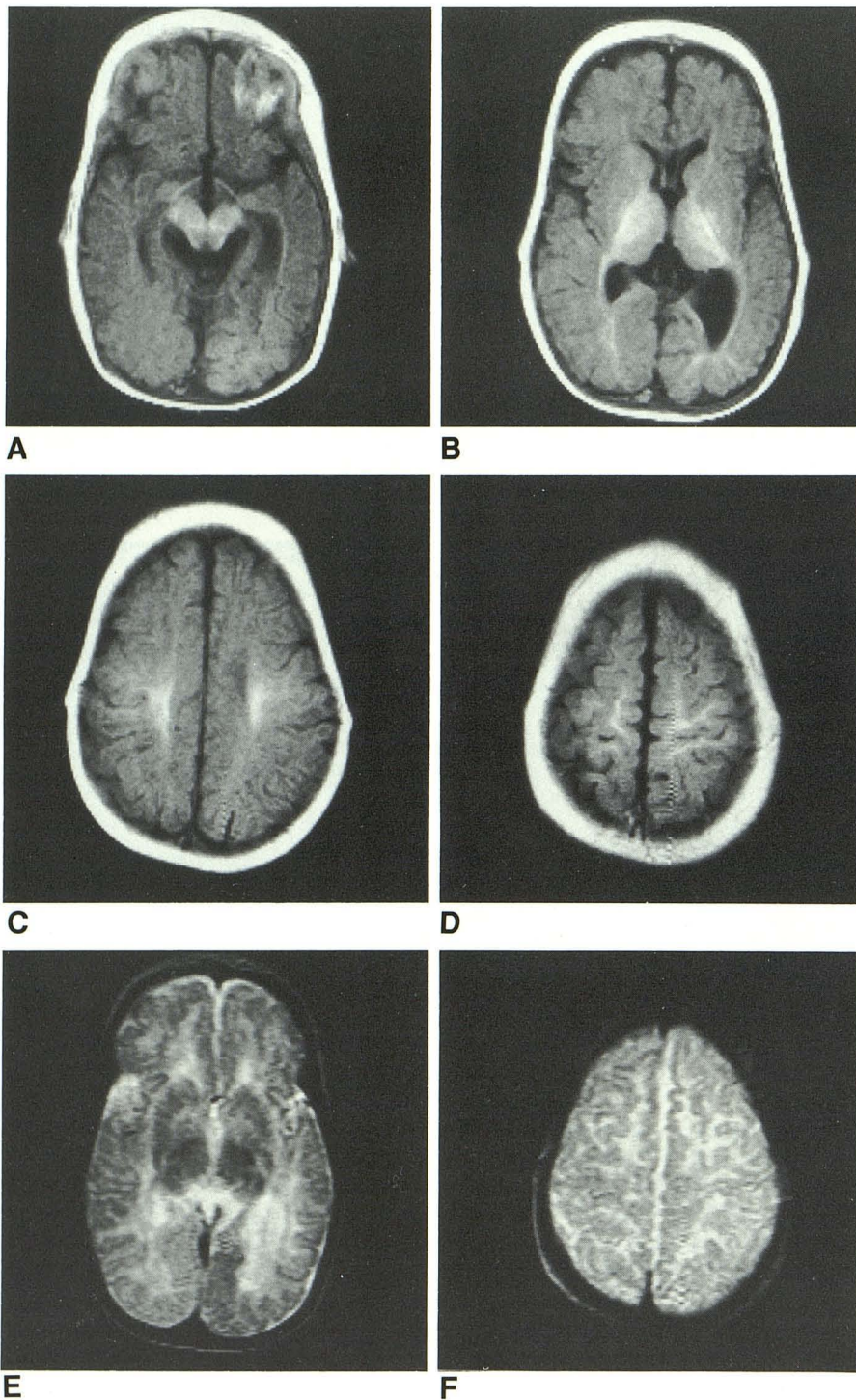


Fig. 8.—Classification: retarded myelination (5.2.).

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-

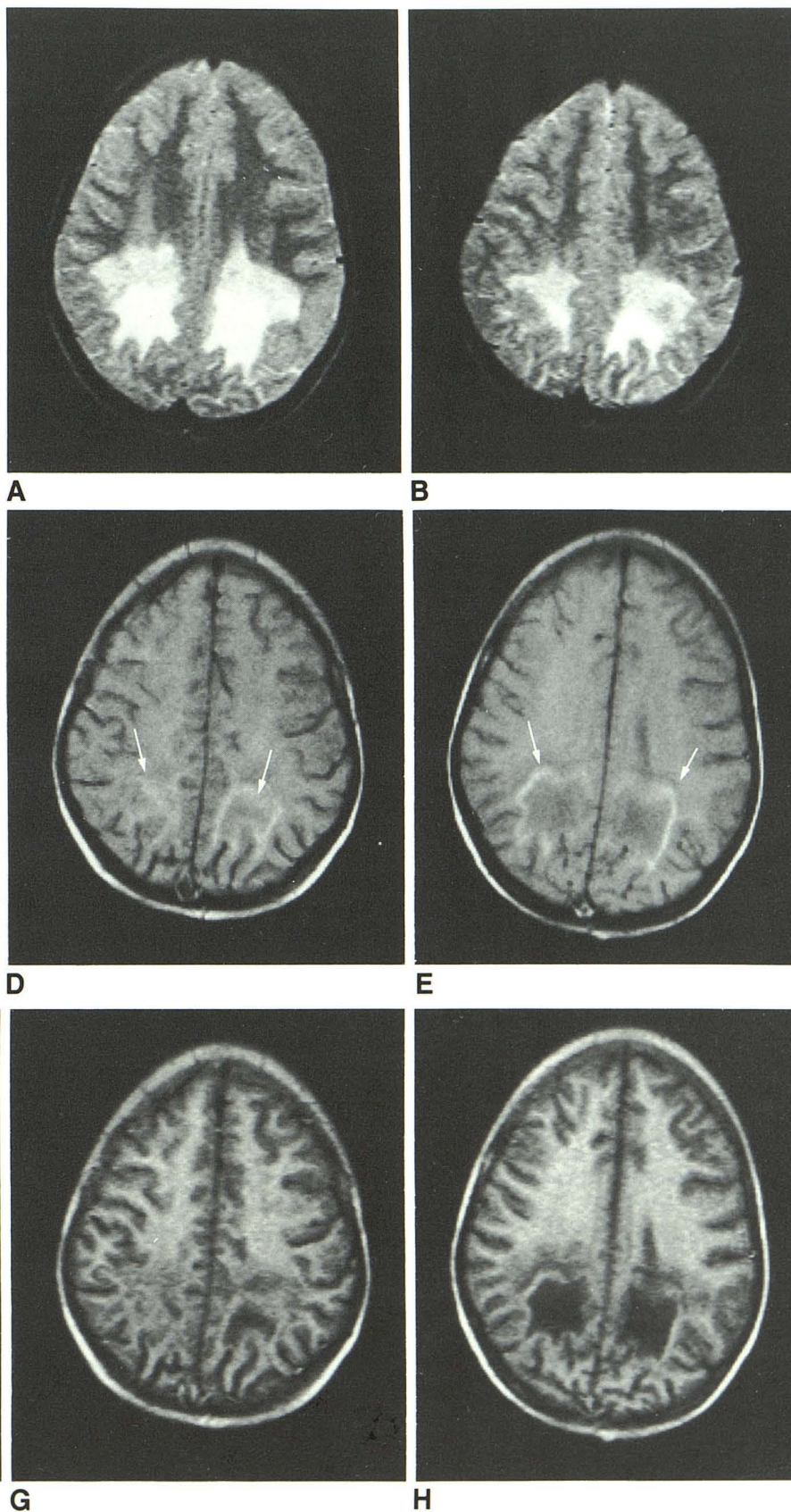
phaly for clefts in the brain, with or without hydrocephalus, with or without fused lips, either unilateral or bilateral, and either developmental or encephaloclastic. Porencephaly is reserved for a cavity within the cerebral hemisphere that does not interconnect the lateral ventricle and the subarachnoid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





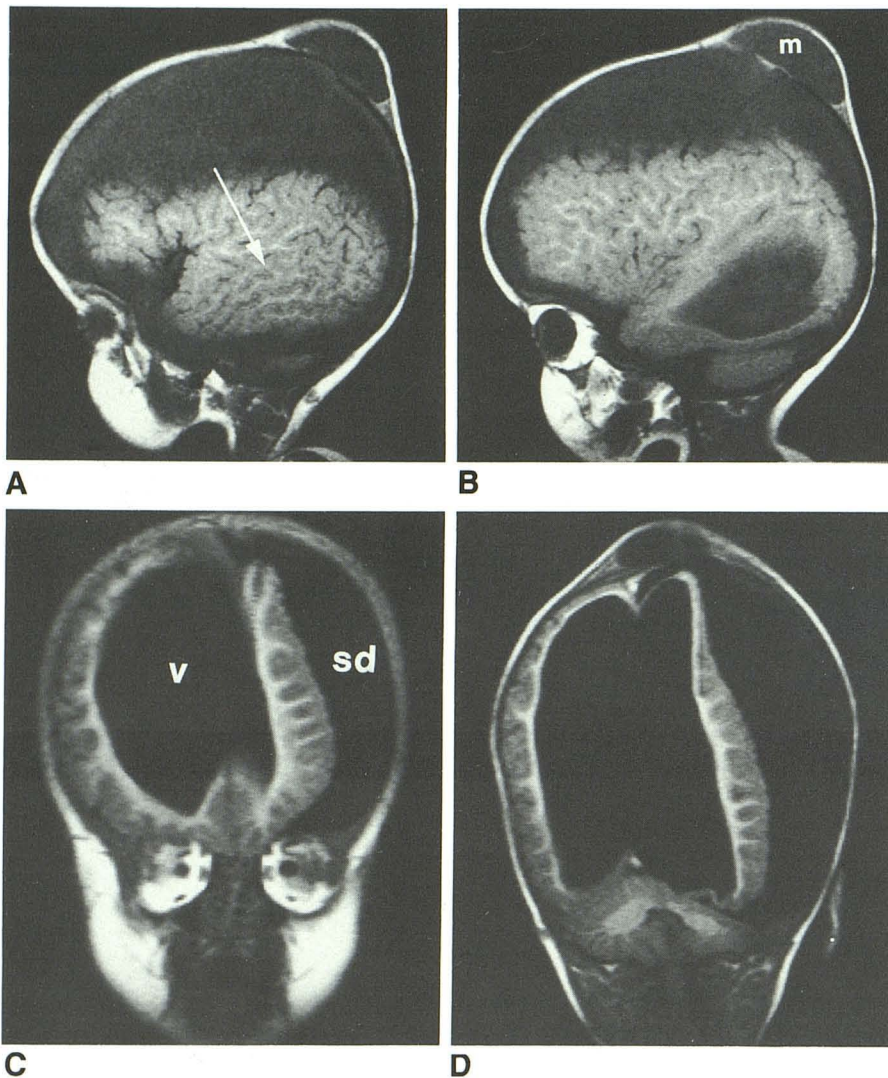


Fig. 10.—Case difficult to classify because it contains elements of more than one subset.

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 reviewing 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.

## REFERENCES

- Johnson MA, Pennock JM, Bydder GM, et al. Clinical NMR imaging of the brain in children: normal and neurologic disease. *AJR* 1983;141:1005–1018
- Volpe JJ. *Neurology of the newborn. Major problems in clinical pediatrics*, vol. 22, 2nd ed. Philadelphia: Saunders, 1987
- Naidich TP, McLone DG. Congenital pathology of the spine and spinal cord. In: Taveras JM, Ferrucci JT, eds. *Radiology: diagnosis-imaging-intervention*, vol. 3. Philadelphia: Lippincott, 1986:1–23
- Dryden RJ. Duplication of the spinal cord: a discussion of the possible embryogenesis of diplomyelia. *Dev Med Child Neurol* 1980;22:234–243
- Saunders RL. Intramedullary epidermoid cyst associated with a dermal sinus. *J Neurosurg* 1969;31:83–86
- Sarnat HB, Case ME, Graviss R. Sacral agenesis. *Neurology* 1976;26:1124–1129
- Ignelzi RJ, Lehman RAW. Lumbrosacral agenesis: management and embryological implications. *J Neurol Neurosurg Psychiatry* 1974;37:1273–1276
- Manelfe C, Sevely A. Neuroradiological study of holoprosencephalies. *J Neuroradiol* 1982;9:15–45
- Hale BR, Rice P. Septo-optic dysplasia: clinical and embryological aspects. *Dev Med Child Neurol* 1974;16:812–820
- Morishima A, Aranoff GS. Syndrome of septo-optic-pituitary dysplasia: the clinical spectrum. *Brain Dev* 1986;8:233–239
- Jellinger K, Gross H. Congenital telencephalic midline defects. *Neuropediatrics* 1973;4:446–452
- Brett EM, ed. *Paediatric neurology*. London: Churchill Livingstone, 1983
- Yokota A, Oota T, Matsukado Y. Dorsal cyst malformations. *Childs Brain* 1984;11:320–341
- Brocklehurst G. Diencephalic cysts. *J Neurosurg* 1973;38:47–51
- Robinson RG. Agenesis and anomalies of other brain structures. In: Vinken PJ, Bruyn GW, eds. *Handbook of clinical neurology*, vol. 30. Amsterdam: North Holland, 1977:352–359
- Lang C, Lehl S, Huk W. A case of bilateral temporal lobe agenesis. *J Neurol Neurosurg Psychiatry* 1981;44:626–630
- Macchi G, Bentivoglio M. Agenesis or hypoplasia of cerebellar structures. In: Vinken PJ, Bruyn GW, eds. *Handbook of clinical neurology*, vol. 30. Amsterdam: North Holland, 1977:367–393
- Mercuri S, Curatolo P, Giuffrè R, Di Lorenzo N. Agenesis of the vermis cerebelli and malformations of the posterior fossa in childhood and adolescence. *Neurochirurgia (Stuttg)* 1979;22:180–188
- Calogero JA. Vermian agenesis and unsegmented midbrain tectum. *J Neurosurg* 1977;47:605–608
- Joubert M, Eisenring JJ, Robb JP, Andermann F. Familial agenesis of the cerebellar vermis. *Neurology* 1969;19:813–825
- Hirsch JF, Pierre-Kahn A, Renier D, Sainte-Rose C, Hoppe-Hirsch E. The Dandy-Walker malformation: a review of 40 cases. *J Neurosurg* 1984;61:515–522
- Gardner E, O'Rahilly R, Prolo D. The Dandy-Walker and Arnold-Chiari malformations: clinical, developmental and teratological considerations. *Arch Neurol* 1975;32:393–407
- Rayband C. Cystic malformations of the posterior fossa. *J Neuroradiol* 1982;9:103–133
- Moss ML. Functional anatomy of cranial synostosis. *Childs Brain* 1975;1:22–33
- Zuleta A, Basauri L. Cloverleaf skull syndrome. *Childs Brain* 1984;11:418–427
- Robain G. Familial micrencephalies due to cerebral malformation. *Acta Neuropathol (Berl)* 1972;20:96–109
- DeMyer W. Megalencephaly in children: clinical syndromes, genetic patterns and differential diagnosis from other causes of megalencephaly. *Neurology* 1972;22:634–643
- Townsend JJ, Nielsen SL, Malamud N. Unilateral megalencephaly: hamartoma or neoplasm? *Neurology* 1975;25:448–453
- Menkes JH. *Textbook of child neurology*. Philadelphia: Lea & Febiger, 1980
- Francois J. A general introduction to the phakomatoses. In: Vinken PJ, Bruyn GW, eds. *Handbook of clinical neurology*, vol. 14. Amsterdam: North Holland, 1972:1–18
- Warkany J, Lemire RJ, Cohen MM. *Mental retardation and congenital malformations of the central nervous system*. Chicago: Year Book Medical, 1981
- Ohta T, Kajikawa H, Takeuchi J. Congenital tumours of the brain. In: Vinken PJ, Bruyn GW, eds. *Handbook of clinical neurology*, vol. 31. Amsterdam: North Holland, 1977:35–74
- Raimondi AJ, Tomita T. Brain tumors during the first year of life. *Childs Brain* 1983;10:193–207
- Faivre J, Lemarec B, Bretagne J, Pecker J. X-linked hydrocephalus with aqueductal stenosis, mental retardation and adduction-flexion deformity of the thumbs. *Childs Brain* 1976;2:226–233
- Garg BP. Colpocephaly: an error of morphogenesis. *Arch Neurol* 1982;39:243–246
- Herskowitz J, Rosman NP, Wheeler CB. Colpocephaly: clinical, radiologic and pathogenetic aspects. *Neurology* 1985;35:1594–1598
- Smit LME, Barth PG, Valk J, Njikiktjen C. Familial porencephalic white matter disease in two generations. *Brain Dev* 1984;6:54–58
- Suarez JC, Sjaello ZM, Albarenque M, Viano JC. Porencephalic congenital cysts with hydrocephalus. *Childs Brain* 1984;11:77–86
- Berg RA, Kyriakos AA, Kaplan AM. Familial porencephaly. *Arch Neurol* 1983;40:567–569
- Naidich TP, Chakera TMH. Multicystic encephalomalacia: CT appearance and pathologic correlation. *J Comput Assist Tomogr* 1984;8:613–616
- Halsey JH, Allen N, Chamberlin HR. The morphogenesis of hydranencephaly. *J Neurol Sci* 1971;12:187–127
- Fowler M, Dow R, White TA, Greer CH. Congenital hydrocephalus-hydrencephaly in five siblings with autopsy studies: a new disease. *Dev Med Child Neurol* 1972;14:173–188
- Yakovlev PI, Wadsworth RC. Schizencephalies: a study of the congenital clefts in the cerebral mantle. I. Clefts with fused lips. *J Neuropathol Exp Neurol* 1946;5:116–130
- Yakovlev PI, Wadsworth RC. Schizencephalies: a study of the congenital clefts in the cerebral mantle. II. Clefts with hydrocephalus and lips separated. *J Neuropathol Exp Neurol* 1946;5:169–206
- Page LK, Brown SB, Gargano FP, Shortz RW. Schizencephaly: a clinical study and review. *Childs Brain* 1975;1:348–358
- Dambska M, Wisniewski K, Sher JH. Lissencephaly: two distinct clinicopathological types. *Brain Dev* 1983;5:302–310
- Hanaway J, Lee SI, Netsky MG. Pachygyria: relation of findings to modern



- embryologic concepts. *Neurology* **1968**;18:791-799
48. Trounce JQ, Fagan DG, Young ID, Levence MI. Disorders of neuronal migration: sonographic features. *Dev Med Child Neurol* **1986**;28:467-471
  49. Davison AN, Dobbing J, Path MC. Myelination as a vulnerable period in brain development. *Br Med Bull* **1966**;22:40-44
  50. Garcia CA, Duncan C. Atelencephalic microcephaly. *Dev Med Child Neurol* **1977**;19:227-232
  51. Crome L. Hydrencephaly. *Dev Med Child Neurol* **1972**;14:224-234
  52. Yoshioka H, Yoshida A, Ochi M, et al. Hydranencephaly in twins. *Brain Dev* **1980**;2:327-329
  53. Schmitt HP. Multicystic encephalopathy—a polyetiologic condition in early infancy: morphologic, pathogenetic and clinical aspects. *Brain Dev* **1984**;6:1-9
  54. Barth PG, Valk J, Olsislagers-de Slegte R. Central cortico-subcortical pattern on CT in cerebral palsy. *J Neuroradiol* **1984**;11:65-71
  55. Grant EG, Williams AL, Schellinger D, Slovis TL. Intracranial calcification in the infant and neonate: evaluation by sonography and CT. *Radiology* **1985**;157:63-68
  56. Tsubokawa T, Nakamura S, Satoh K. Effect of temporary subdural-peritoneal shunt on subdural effusion with subarachnoid effusion. *Childs Brain* **1984**;11:47-59
  57. Rengachary SS, Watanabe I, Brackett CE. Pathogenesis of intracranial arachnoid cysts. *Surg Neurol* **1978**;9:139-144
  58. Rengachary SS, Watanabe I. Ultrastructure and pathogenesis of intracranial arachnoid cyst. *J Neuropathol Exp Neurol* **1981**;40:61-83
  59. Giudicelli G, Hassoun J, Choux M, Tonon C. Supratentorial arachnoid cysts. *J Neuroradiol* **1982**;9:179-201
  60. Moore KL. *The developing human*, 2d ed. Philadelphia: Saunders: **1977**
  61. Hamilton WJ, Mossman HW. *Hamilton, Boyd and Mossman's human embryology. Prenatal development of form and function*, 4th ed. Cambridge: Heffer, **1972**
  62. Barry A, Patten BM, Stewart BH. Possible factors in the development of the Arnold-Chiari malformation. *J Neurosurg* **1957**;14:285-301
  63. Salam MZ, Adam SRD. The Arnold-Chiari malformation. In: Vinken PJ, Bruyn GW, eds. *Handbook of clinical neurology*, vol. 32. Amsterdam: North Holland, **1977**:99-110
  64. Schliep G. Syringomyelia and syringobulbia. In: Vinken PJ, Bruyn GW, eds. *Handbook of clinical neurology*, vol. 32. Amsterdam: North Holland, **1977**:255-327
  65. Warkany J, O'Toole BA. Experimental spina bifida and associated malformations. *Childs Brain* **1981**;8:18-30
  66. Frerebeau P, Dimeglio A, Gras M, Harbi H. Diastematomyelia: report of 21 cases surgically treated by a neurosurgical and orthopedic team. *Childs Brain* **1983**;10:328-339
  67. Norman RM. Malformations of the nervous system, birth injury and diseases of early life. In: Greenfield JG, ed. *Neuropathology*. London: Edward Arnold Ltd, **1967**
  68. Dale AJD. Diastematomyelia. *Arch Neurol* **1969**;20:309-317
  69. Carter CO, Evans KA, Till K. Spinal dysraphism: genetic relation to neural tube malformations. *J Med Genet* **1976**;13:343
  70. Lee BCP, Lipper E, Nass R, Ehrlich ME, de Ciccio-Bloom E, Auld PAM. MRI of the central nervous system in neonates and young children. *AJNR* **1986**;7:605-616
  71. Holland BA, Haas DK, Norman D, Brant-Zawadzki M, Newton TH. MRI of normal brain maturation. *AJNR* **1986**;7:201-208
  72. McArdle CB, Richardson CJ, Nicholas DA, Mirfakhraee M, Hayden CK, Ampara EG. Developmental features of the neonatal brain: MR imaging. *Radiology* **1987**;162:223-229
  73. Raybaud C. Destructive lesions of the brain. *Neuroradiology* **1983**;25:265-291