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Brain MR in Fukuyama Congenital Muscular Dystrophy

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PURPOSE: To determine the MR characteristics of brain abnormalities in Fukuyama congenital muscular dystrophy (FCMD). **METHODS:** We reviewed 30 MR examinations of 21 patients with FCMD to assess cerebral and cerebellar cortical dysplasia, white matter changes, and miscellaneous abnormalities. **RESULTS:** On MR images, all patients had thick and bumpy cortices with shallow sulci corresponding to polymicrogyria, and 12 patients had pachygyric cortices with smooth surfaces, corresponding to type II lissencephaly. Both types of cortical dysplasia had characteristic distributions: the first type involved the frontal lobe in all 21 patients and also the parietotemporal lobe in 6 patients; the second type involved the temporooccipital lobes. Eighteen patients had prolonged T1 and T2 signal in the white matter, which was indistinct in neonates and seen infrequently in adolescents. In four patients, abnormal vessels were seen within the pachygyric cortices. **CONCLUSION:** MR studies of the brain show findings consistent with the known characteristics of FCMD. The MR detection of the two types of cerebral cortical dysplasia with characteristic distribution and cerebellar abnormalities is helpful in the differential and early diagnosis of FCMD.

Index terms: Brain, magnetic resonance; Children, diseases; Muscular dystrophy

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Fukuyama congenital muscular dystrophy (FCMD) is the second most common form of muscular dystrophy in Japan, exceeded only by Duchenne muscular dystrophy; however, it is rare in other countries (1). It is a transmitted autosomal recessive trait (2, 3) and the gene locus has recently been mapped on chromosome 9 (4). The clinical features include muscular hypotonia with prenatal onset and severe developmental delay. The diagnosis is based on both the clinical presentation and the pathologic evidence of muscular dystrophy on biopsy specimens. In addition to the histologic change

that occurs in the muscle, FCMD is accompanied by a variety of neuropathologic abnormalities, including cerebral and cerebellar cortical dysplasia (2, 3, 5). These cerebral changes have been intensively investigated by Takada et al (6, 7), who noted three patterns of cerebral cortical dysplasia: verrucous dysplasia (type 1), unlayered polymicrogyria (type 2), and a pattern that is essentially identical to type II lissencephaly (type 3). These authors have also described the characteristic topographic predilection evidenced by each type of dysplasia: the medial surface of the occipital lobe for type 1, the frontal and parietal lobe for type 2, and the lateral surface of the occipital and temporal lobes for type 3 (6, 7). The cerebellum is characterized by polymicrogyria, mainly in the superior semilunar lobule (6, 8). Despite the dearth of neuropathologic descriptions, the abnormality of the white matter has been a focus of the radiologic literature pertaining to FCMD (2, 3, 9, 10). The purpose of this study was to determine the magnetic resonance (MR) characteristics and distribution of brain abnormalities in FCMD.

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Subjects and Methods

Twenty-one patients with FCMD in whom diagnoses had been made on the basis of clinical features and muscle biopsy findings had a total of 30 MR studies available for assessment. At the time of imaging examination, the patients (12 male and 9 female) ranged in age from 10 days to 19 years. Twenty-nine of the 30 studies were performed with a 1.5-T scanner and one was performed with a 0.5-T scanner. Spin-echo T1-weighted images (400–450/15–20/2 [repetition time/echo time/excitations]), T2-weighted images (2500–3000/80–120/1), and fast spin-echo T2-weighted images (5000/112/2; echo train, 15) were obtained.

We evaluated the MR imaging results to ascertain the type, frequency, and distribution of abnormalities, including cerebral and cerebellar cortical dysplasia and white matter disorders. We also determined the prevalence of enlarged subarachnoid spaces, ventriculomegaly, abnormalities of the brain stem, and miscellaneous abnormalities.

Results

The MR findings for all 21 patients are summarized in the Table.

Cerebral Cortical Dysplasia

MR imaging depicted two patterns of cerebral cortical dysplasia. Both patterns appeared to be symmetrical and their frequencies showed no relation to the patients' ages. The first pattern was that of a slightly thickened cortex with shallow sulci and a bumpy gray–white matter interface (Figs 1–4). This pattern was recognized in all 21 patients and involved the frontal lobe in all and the temporoparietal lobes in a subgroup of 6 patients. The MR imaging characteristics of this pattern were typical of polymicrogyria. In some patients the sagittal images made it easier to detect the changes of this cortical dysplasia that were limited to the frontal lobe (Fig 3B–E).

The second pattern was characterized by a thick cortex with a smooth surface and a smooth gray–white matter interface (Fig 1). This was found in the temporooccipital lobes of 12 of the 21 patients. The cortex identified in the second MR pattern was thicker than that in the first MR pattern, and was typical of pachygyria. Six of 12 patients who had the pachygyric pattern, most of whom were younger than 3 years old, had curvilinear bands of increased T2 signal deep within the abnormal cortices (Fig 1C and D). One atypical finding occurred in a 9-year-old girl (patient 16) who had extremely severe white matter abnormality.

Cerebellar Malformation

As we reported previously (8), disorganized cerebellar foliation corresponding to polymicrogyria with intermingled islands of granular and molecular layers was seen in 18 of our 21 patients, and intraparenchymal cysts located in the peripheral hemispheres closely related to the polymicrogyria were seen in 19 patients (Figs 2A and D and 3A and D). The first pattern was seen most clearly on T2-weighted images, while the second pattern was clearly recognized on all imaging sequences. Both types of abnormalities could be seen regardless of the patients' ages, except for one patient (patient 1) in whom cerebellar cysts, not detected at 10 days of age, were seen at the follow-up examination at 11 months of age. There was no obvious relationship between the extent or type of cerebral and cerebellar abnormalities.

White Matter Abnormality

Eighteen patients had prolonged T1 and T2 signal in the white matter (Figs 1–4). The signal alteration was variable in extent among patients and with age. The signal alteration of the white matter was widespread on all 9 MR studies obtained in late infancy and early childhood (6 months to 21 months), whereas among the 14 studies obtained between the ages of 34 months and 19 years, it was diffuse on 3, patchy and spotty on 9, and not recognized on 2. The signal change was more difficult to detect in neonates and infants. MR studies in 2 neonates (patients 1 and 2) and a 5-month-old boy (patient 4) failed to show the signal alteration of the white matter (Fig 4 A and B); follow-up MR studies obtained in late infancy showed diffuse T1 and T2 prolongation in patients 1 and 2 (Fig 4 C and D); and follow-up MR studies were not done in patient 4.

The abnormality of the white matter showed a peculiar pattern of evolution; in three patients (patients 3, 5, and 7), who had diffuse T1 and T2 prolongation of the white matter in infancy, T1 and/or T2 shortening ensued with time in the parietooccipital subcortical region and in the corpus callosum (Fig 1C and D); in another patient (patient 9), patchy T2 prolongation of the white matter evolved into spotty lesions during childhood. The clinical status of six patients, in whom serial MR examinations were per-

MR findings in 21 patients with Fukuyama congenital muscular dystrophy

Patient	Sex/Age	Cerebral Cortical Dysplasia				Cerebellar Abnormality		White Matter Abnormality	Hypoplasia of Brain Stem	Ventriculomegaly	Enlarged Subarachnoid Space
		PMG	PG	Band	Vessel	PMG	Cyst				
1	F/10 d	Frontal	Temporal	-	-	-	-	-	±	-	-
	11 mo	Frontal	Temporal	+	-	+	+	Diffuse	+	-	-
2	M/19 d	Frontal	Temporal	-	-	+	+	-	±	-	-
	2 mo	Frontal	Temporal	-	-	+	+	Equivocal	±	-	-
	6 mo	Frontal, parietal	Temporal	-	-	+	+	Diffuse	+	-	-
3*	F/2 mo	Frontal, parietal	Temporal, occipital	-	-	+	+	Diffuse	+	+	+
	3 mo	Frontal, parietal	Temporal, occipital	-	-	+	+	Diffuse	+	+	+
	9 mo	Frontal, parietal	Temporal, occipital	-	-	+	+	Diffuse, T1 area	+	+	+
4*	M/5 mo	Frontal	-	-	+	-	-	+	+
5*	M/5 mo	Frontal, parietal	Temporal, occipital	+	-	+	+	Diffuse	+	+	+
	21 mo	Frontal, parietal	Temporal, occipital	+	+	+	+	Diffuse, T1, T2 area	+	+	+
6*	M/7 mo	Frontal	Temporal, occipital	+	-	+	+	Diffuse	+	+	+
7*	F/8 mo	Frontal, parietal, temporal	-	+	+	Diffuse	+	+	+
	11 mo	Frontal, parietal, temporal	-	+	+	Diffuse, T1, T2 area	+	+	+
	20 mo	Frontal, parietal, temporal	-	+	+	Diffuse, T1, T2 area	+	+	+
8	F/14 mo	Frontal	-	+	+	Diffuse	+	-	-
9*	F/34 mo	Frontal	-	+	+	Patchy	-	-	-
	5 y	Frontal	-	+	+	Spotty	-	-	-
10	M/35 mo	Frontal, parietal	Temporal, occipital	+	+	+	+	Patchy	+	+	+
11	M/5 y	Frontal	Temporal, occipital	-	+	+	+	Diffuse	+	+	+
12	M/6 y	Frontal	-	+	+	-	+	±	-
13*	M/6 y	Frontal	Temporal, occipital	-	-	+	+	Patchy	+	+	+
14	M/7 y	Frontal	Temporal, occipital	-	+	+	+	Diffuse	+	+	+
15	M/9 y	Frontal, temporal	-	+	+	Patchy	+	+	+
16*	F/9 y	Frontal	Occipital	+	-	-	-	Diffuse	-	+	+
17	F/11 y	Frontal	Temporal, occipital	-	-	+	+	Patchy	+	+	+
18	M/15 y	Frontal	-	-	-	Patchy	-	+	+
19*	M/16 y	Frontal	Temporal	-	-	+	+	Spotty	+	+	+
20*	M/17 y	Frontal	-	+	+	-	-	+	+
21*	F/19 y	Frontal	-	+	+	Spotty	-	+	+

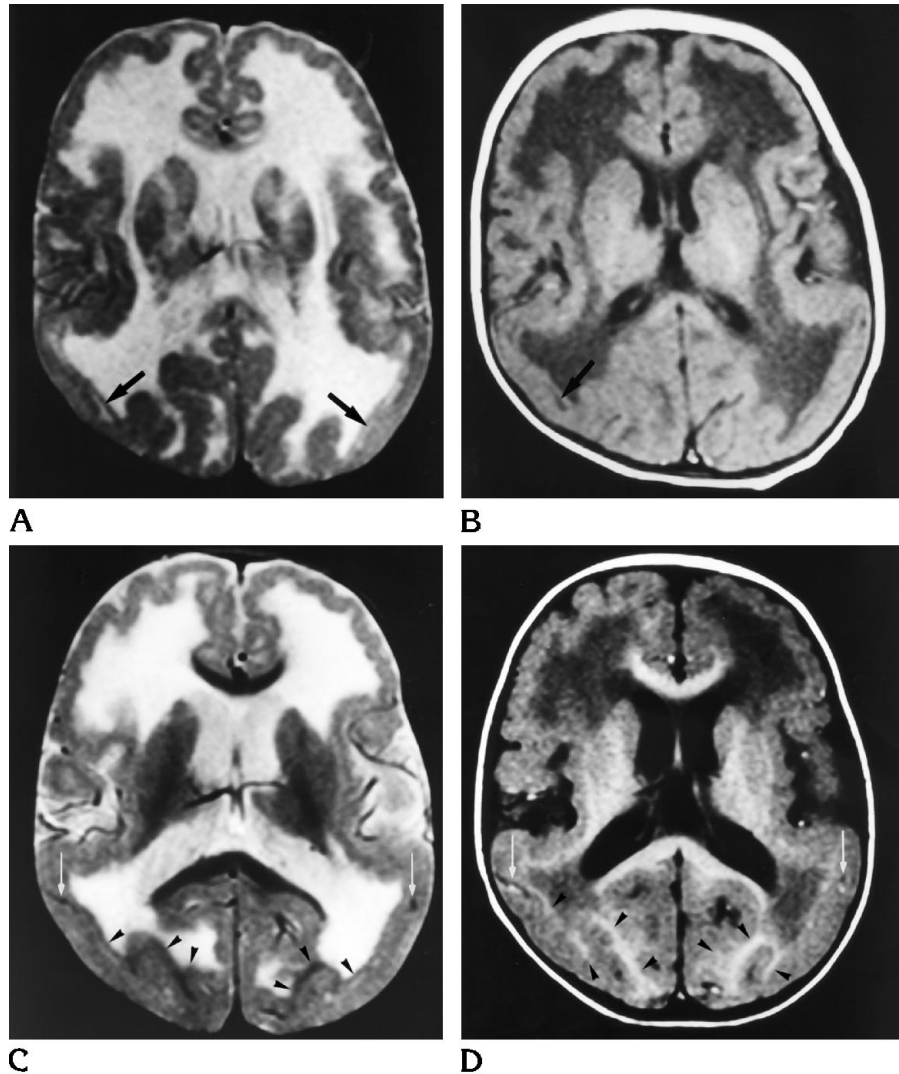
Note.—PMG indicates polymicrogyria, Takada type 2 in the cerebrum; PG, pachygyria, Takada type 3; band, curvilinear band of increased T2 signal; vessel, abnormal vessels buried beneath the area of pachygyric cortical dysplasia; T1 and T2 area, areas of T1 or T2 shortening (hyperintense on T1-weighted images and hypointense on T2-weighted images) were apparent; +, present; and -, absent. Patient 6 is a sibling of patient 1; patient 17 is a sibling of patient 14.

* Cerebellar findings from these patients have been published previously (8).

Fig 1. Patient 5.

A, T2-weighted and B, T1-weighted axial MR images of a 5-month-old boy. The frontal lobes show slightly thick and bumpy cortices with shallow sulci (polymicrogyria), and the temporooccipital lobes show pachygyric cortices with smooth surfaces. Curvilinear bands of increased T1 and T2 signal (arrows) are present deep within the pachygyric cortices, and diffuse prolonged T1 and T2 signal is present in the white matter.

C, T2-weighted and D, T1-weighted axial MR images obtained at the follow-up study at age 1 year 9 months. Note shortening of T1 and T2 signal in the corpus callosum and the occipital subcortical region (arrowheads) caused by progressive myelination. Some abnormally located vessels are seen within the pachygyric cortices (arrows). The lateral ventricles are slightly enlarged.



formed between the ages of 10 days and 5 years, improved very slowly.

Miscellaneous Abnormalities

Abnormal cortical vessels were buried beneath the area of pachygyric cortical dysplasia in 4 of 12 patients (Fig 1 C and D). However, this vascular abnormality was not seen within polymicrogyric cortices. An enlarged subarachnoid space with ventriculomegaly of varying degrees was seen on MR images in 17 patients (Figs 1 and 3). These findings did not change on serial studies. Flattening of the ventral portion of the pons at the level of the middle cerebellar peduncle (Fig 3A) was detected in 15 patients.

Discussion

Congenital muscular dystrophy (CMD) is a heterogeneous group of disorders characterized by muscular hypotonia of prenatal onset and the histologic features of muscular dystrophy. It is classified on the basis of the clinical features, pathologic findings, and pattern of inheritance. An association of CMD and migration disorder is found in FCMD, Walker-Warburg syndrome, and muscle-eye-brain disease (2, 3, 5-8, 11-15). The classification of these disorders remains controversial. In Japan, FCMD is the most common congenital muscular dystrophy, and there are also reports from Australia and the Netherlands of non-Japanese patients with FCMD (16-19). Thus, it is important for all radiologists to be aware of this entity, even though it is uncommon outside Japan.

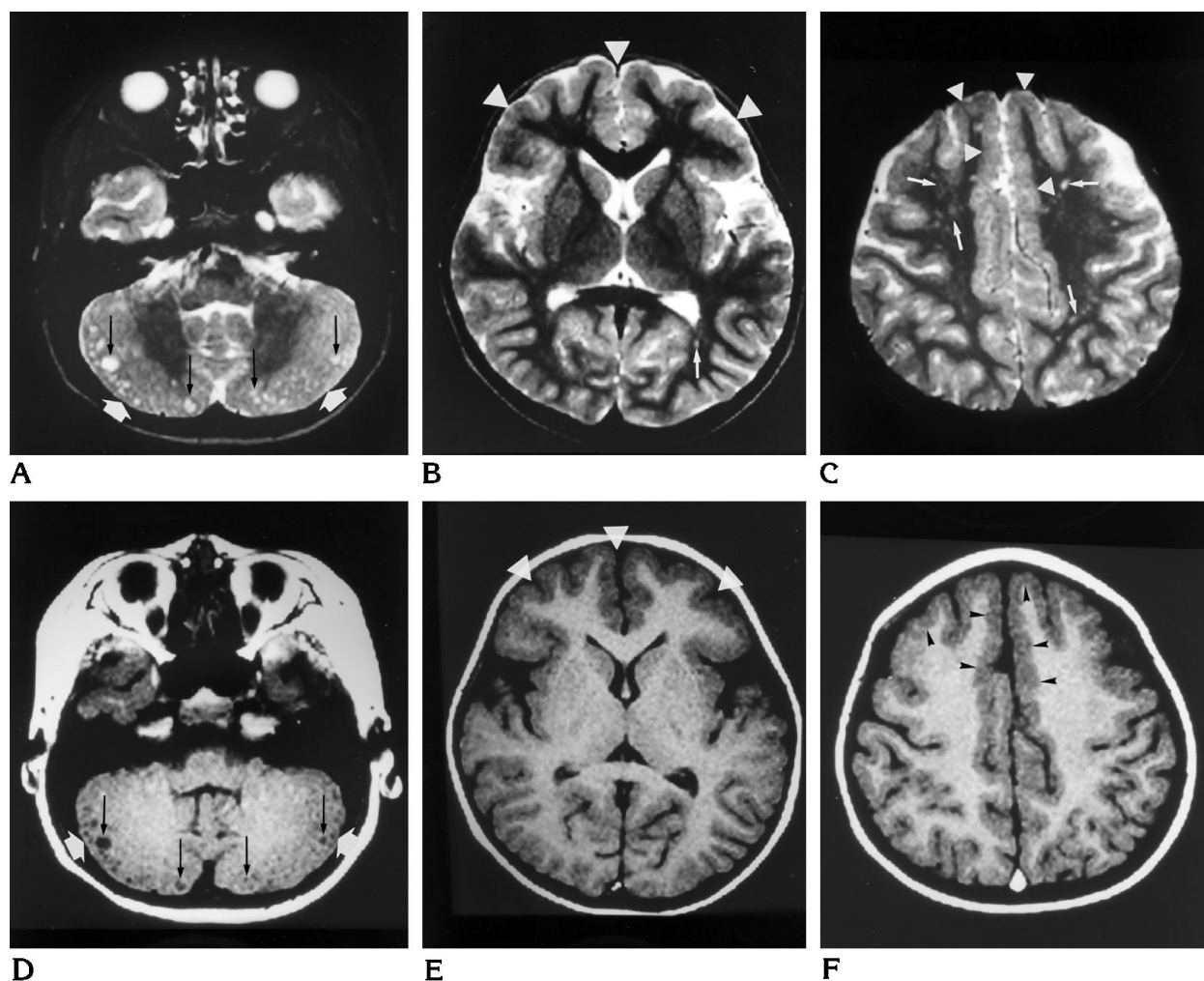


Fig 2. Patient 9.

T2-weighted (A) and T1-weighted (D) axial MR images of the middle portion of the cerebellum in a 5-year-old girl show diffuse disorganized cerebellar folia (cerebellar polymicrogyria; *white arrows*) with numerous intraparenchymal cysts (*black arrows*) at the peripheral hemispheres. T2-weighted (B and C) and T1-weighted (E and F) axial MR images of the cerebrum show slightly thick and bumpy cortices with abnormal gyration (*arrowheads*) in the frontal lobes. The temporooccipital cortices show no abnormality. The white matter change is minimal (*arrows*).

FCMD causes severe mental retardation, seizures, and muscular weakness soon after birth, and it is associated with a variety of abnormalities of the central nervous system (2, 3, 6–8). The diagnosis is classically based on pathologic evidence of muscular dystrophy in the biopsy specimen in the appropriate clinical context. However, neurologic examination is not easy and even the histologic features occasionally may be ambiguous, particularly in neonates.

Cerebral Cortical Dysplasia

The histopathologic features of cerebral abnormalities in FCMD have been investigated by

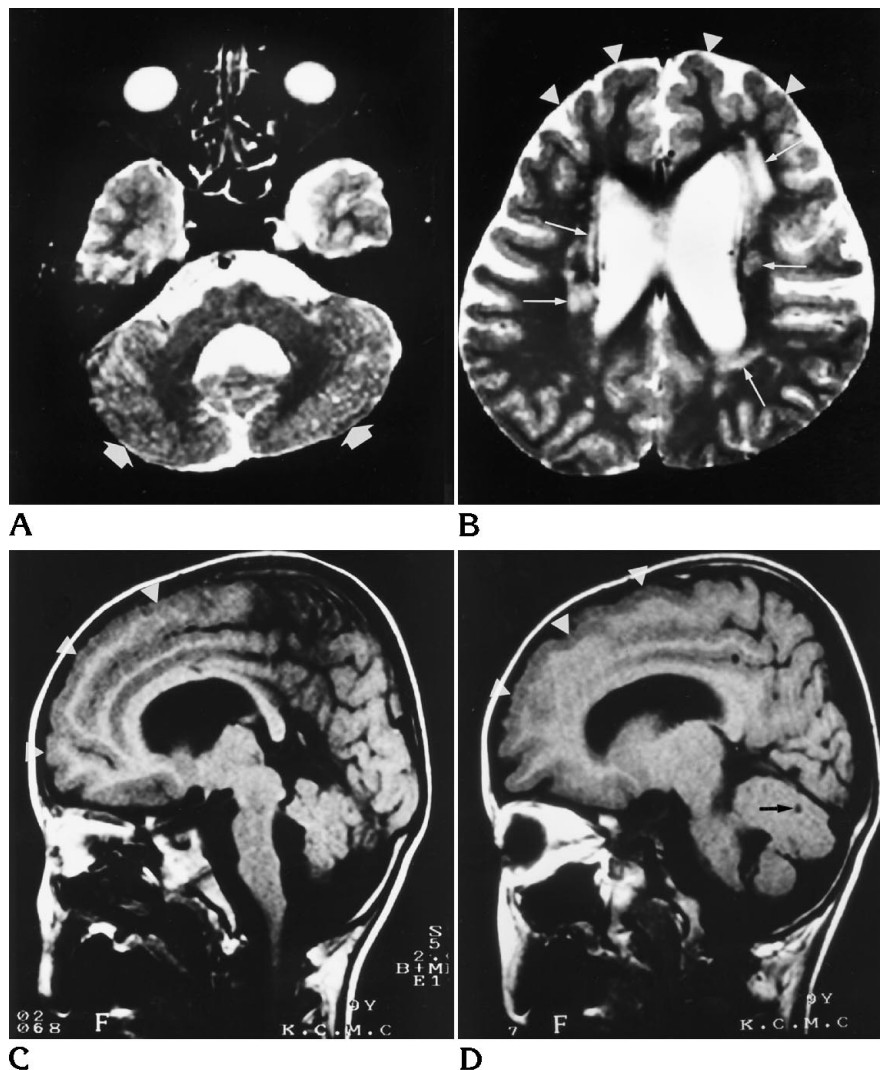
Takada et al (6, 7), who summarized them as follows: verrucous dysplasia with cortical nodules, characterized microscopically by an elevated cellular bump with absence of the molecular layer (type 1); unlayered polymicrogyria, characterized microscopically by a single, thick cellular layer separated by numerous narrow acellular zones (microsulci) (type 2); and, the most severe type of cortical dysplasia (type 3), which is essentially identical to type II lissencephaly, characterized macroscopically by completely smooth surfaces and four distinctive layers consisting of the superficial molecular layer containing tangential myelinated fibers, a thick cellular layer, a layer composed of a large

Fig 3. Patient 15.

A, T2-weighted axial MR image of the middle portion of the cerebellum in a 9-year-old boy reveals disorganized cerebellar folia (cerebellar polymicrogyria; *arrows*), flattening of the ventral portion of the pons, and an enlarged fourth ventricle.

B, T2-weighted axial MR image of the cerebrum shows a subtle polymicrogyric change in the frontal lobe (*arrowheads*), appearing as shallow sulci. Note the enlarged subarachnoid space, ventriculomegaly, and the few periventricular areas of increased T2 signal lesions (*arrows*).

C and D, Sagittal T1-weighted MR images reveal abnormal cortical architecture of the frontal lobe (*arrowheads*) as evidenced by shallow sulci and a bizarre pattern of the sulci parallel to each other without perpendicular sulci. This contrasts with the relatively normal parietooccipital sulcation. The cerebellum shows abnormal folia with a small cyst (*arrow*). A flattened pons and enlargement of the fourth ventricle are conspicuous.



number of myelinated fibers, and the deep cellular layer (6). These authors also described the characteristic regional distribution for each type of cortical dysplasia: for type 1, it is the medial surface of the occipital lobe; for type 2, the frontal and parietal lobes; and for type 3, the lateral surface of the occipital and temporal lobes.

In our series, MR imaging demonstrated two types of cerebral cortical dysplasia that correspond to the known pathologic patterns of FCMD. We believe that the MR pattern of a thick cortex with shallow sulci and a bumpy gray-white matter interface involving the frontal lobes that we observed in all our patients is typical of polymicrogyria and corresponds to type 2 dysplasia (unlayered polymicrogyria). The second MR pattern of a thick cortex with a smooth surface found in the temporooccipital lobes (Fig 1)

was typical of pachygyria and likely to reflect type 3 dysplasia (type II lissencephaly). We did not recognize any subtle MR abnormalities that might correspond to type 1 dysplasia, probably because of insufficient spatial resolution. MR imaging thus may be used to demonstrate the two types of cerebral cortical dysplasia (type 2 and 3 cortical dysplasia) with their characteristic distributions. These cerebral cortical dysplasias may be associated with several conditions, including other entities classed as CMD, such as Walker-Warburg syndrome and muscle-eye-brain disease (11-13, 20). However, to our knowledge, no such peculiar distribution of cortical dysplasia as described above has been reported in any other condition. These MR changes of the cerebral cortex were seen in all of our patients, regardless of age, indicating that these findings may assist in the diagnosis of

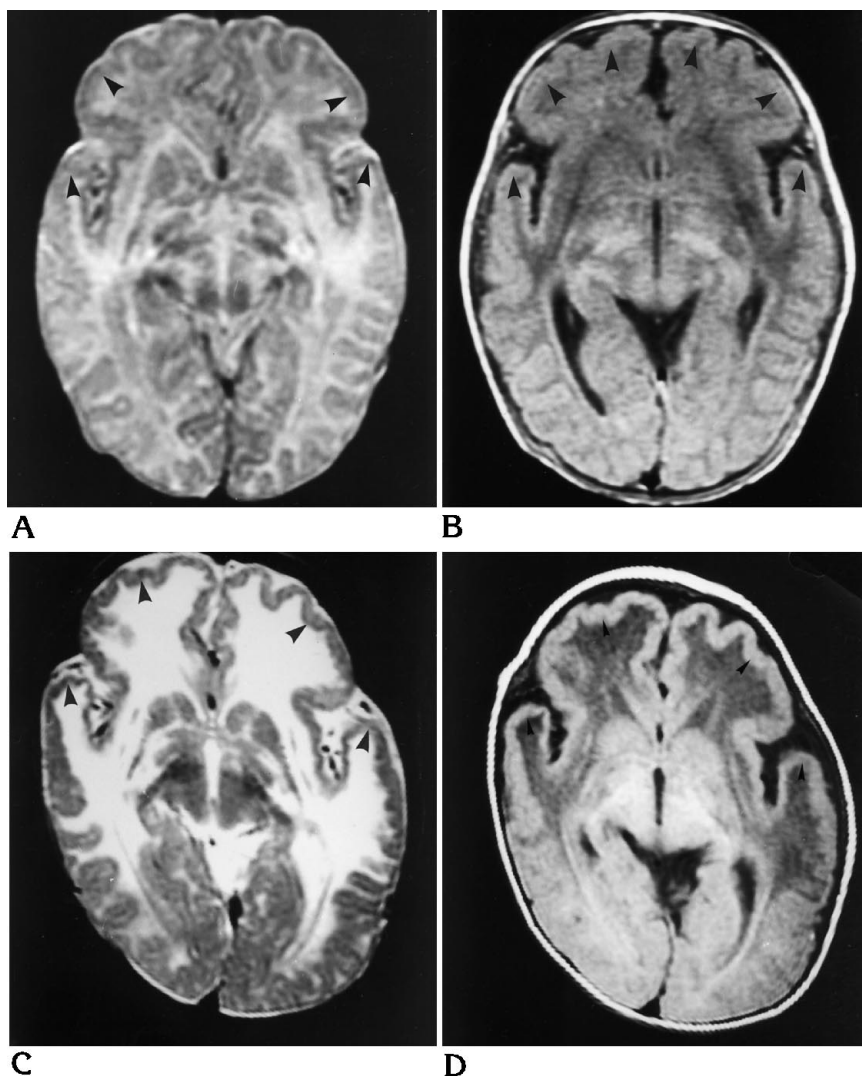


Fig 4. Patient 2.

A, T2-weighted and B, T1-weighted axial MR images in a 19-day-old neonate reveal areas of cerebral cortical dysplasia in the frontal and temporal lobes (*arrowheads*). There is no definite white matter change.

C, T2-weighted and D, T1-weighted axial MR images obtained at the follow-up study at the age of 6 months reveal extensive white matter abnormality as well as fronto-temporal cortical dysplasia (*arrowheads*).

FCMD, although a mild abnormality limited to the frontal lobe may be overlooked (Figs 2 and 3).

Half of the patients with pachygyric cortical dysplasia had extensive prolonged T2 signal in the white matter and curvilinear bands of increased T2 signal deep within the abnormal cortices (Fig 1). Although type I lissencephaly (Miller-Dieker syndrome) also displays similar increased T2 signals in the abnormal cortices (21), it can be differentiated from type 3 dysplasia in FCMD by its superficial location. The third of the four distinctive layers in type 3 dysplasia consists of myelinated fibers pathologically (5, 6), and the curvilinear bands of increased T2 signal may correspond to the third layer of fibers before myelination. It is suggested that the myelinated fibers in type 3 dys-

plasia cannot be detected, but the fibers can be seen before myelination.

Cerebellar Malformation

The previously reported cerebellar MR findings in FCMD, consisting of polymicrogyria and accompanying cysts (8), were seen in more than 90% of the patients in this study, regardless of age. The pathologic study reveals that the cerebellar cysts exist within or near polymicrogyria, which usually fuse with and obstruct the sulci in their superficial parts, and that the cysts are partially lined by leptomenigeal tissue and surround a nearly normal molecular layer (8). The cysts are thus likely to have formed from the subarachnoid spaces that were engulfed by the folia of the malformed cortex, particularly in

the boundary between the normal and polymicrogyric cortices (8). We could not find any obvious relationship between the extent or type of cerebral abnormalities and that of the cerebellar abnormalities. Some conditions, such as Walker-Warburg syndrome and congenital infection, can cause cerebellar polymicrogyria (20), and these patients can have similar neuroradiologic findings, although there are no reports to our knowledge of the radiologic findings. Recently, small multiple lesions with high signal on T2-weighted images and low signal on T1-weighted images in the cerebellar hemispheres, which might have a pathogenesis similar to the cysts in FCMD, have been reported in patients with muscle-eye-brain disease (14). However, we think that the cerebellar polymicrogyria with accompanying cysts is an important finding in FCMD, because it can be recognized more readily than mild frontal polymicrogyria (type 2 dysplasia) and should prompt a careful search for the characteristic cerebral cortical abnormalities that were seen in all patients in this study.

White Matter Abnormality

White matter abnormality is a well-known radiologic finding in FCMD as well as in Walker-Warburg syndrome and muscle-eye-brain disease (2, 3, 11–14). The neuroradiologic literature on FCMD has focused primarily on white matter abnormalities, which have been attributed to delayed myelination (9, 10). We do not agree with this explanation, because, in three of our patients, the evolution of the abnormal white matter signal started in the parietooccipital subcortical region, unlike the centrifugal progression of the normal process of myelination. Therefore, we do not believe that the signal alteration of the white matter simply represents delayed myelination. The cause of the white matter abnormalities in FCMD is not clearly elucidated, even in the pathologic literature, in which only myelin pallor and mild gliosis are briefly described (2, 3, 6). The clarification of the nature of the white matter changes in FCMD will require further neuroradiologic and neuropathologic investigation.

The white matter abnormalities appeared to lessen with age; however, we are unable to identify any specific relationship between the clinical status and the white matter changes because of the small number of patients, their

young age, and the relatively short follow-up periods.

Miscellaneous Abnormalities

We found some vessels traversing the parenchyma with type 3 dysplasia in four patients (Fig 1C and D). Takada et al (7, 22) speculated that the pial-glial barrier, which normally acts as a major impediment to migrating neurons, may be disrupted in FCMD, leading to the development of neuroglial heterotopia within the extracortical glial layer. Therefore, the presence of these abnormal vessels within the malformed cortices is essential evidence supporting their hypothesis regarding the pathogenesis of FCMD, because these vessels, which normally traverse the surface of the brain, may have been engulfed in the course of the abnormal development of neuroglia through a defective barrier. However, this finding was seen in less than 20% of patients, probably because MR imaging does not detect vessels of small caliber.

Many patients had an enlarged subarachnoid space, ventriculomegaly, and flattening of the pons. These changes might be attributed to hypoplasia rather than to atrophy of the brain, because the serial studies revealed no distinct progression of these findings.

In summary, we believe that the MR findings described here, particularly the cerebral cortical dysplasia and cerebellar abnormality, are important in establishing the diagnosis of FCMD. In Japan, FCMD is one of the prime diagnostic considerations in “floppy” infants. The diagnosis of FCMD has traditionally depended on the clinical symptoms and on the findings at muscle biopsy. Nevertheless, it is not always easy to evaluate the neurologic status in the young infant, and the muscle biopsy specimen does not provide us with any specific evidence of FCMD beyond that which simply permits the diagnosis of muscular dystrophy. The MR findings reported here are therefore useful for the early diagnosis of FCMD and may facilitate in the differentiation of FCMD from other forms of CMD.

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