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Congenital Ocular Motor Apraxia: Imaging Findings

Michael A. Sargent, Kenneth J. Poskitt, and James E. Jan

PURPOSE: To determine the frequency of cerebellar and cerebral abnormalities on brain imaging studies in children with congenital ocular motor apraxia. **METHODS:** Brain imaging studies were performed in 19 children with typical congenital ocular motor apraxia who were in the care of a visual impairment program at a children's hospital. Independent clinical review categorized the subjects as having partial (n = 10) or expanded (n = 9) congenital ocular motor apraxia on the basis of extent of associated speech or neurodevelopmental problems. Fifteen CT studies and 13 MR examinations of the brain performed in these children were reviewed independently by two pediatric neuroradiologists. Radiologic findings were agreed on by consensus. **RESULTS:** Cerebellar abnormalities were found in 12 of 19 cases. The cerebellar vermis was small in 10 children. A small cerebellar vermis was the only abnormality in five of 10 children with partial congenital ocular motor apraxia and in two of nine children with expanded congenital ocular motor apraxia. Among seven children with a small vermis examined with high-resolution MR imaging, the inferior portion of the vermis was preferentially involved in each case. Of these seven subjects, none of four with partial congenital ocular motor apraxia but two of three with expanded congenital ocular motor apraxia had an abnormality of the superior portion of the vermis. Miscellaneous supratentorial lesions affecting both gray and white matter were found in six subjects. Five of the 19 children had normal imaging findings. **CONCLUSION:** Inferior vermian hypoplasia is the most common abnormality in children with congenital ocular motor apraxia.

Index terms: Eyes, diseases; Cerebellum, vermis; Infants, eyes and orbits

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Gaze and eye movements are controlled at multiple levels within the brain, including the visual and parietal cortex, frontal lobes, and midbrain (1). Recent evidence suggests that the cerebellar vermis is also involved in the control of eye movements (2, 3). Saccades are rapid eye movements that bring the image of interest over the fovea. Pursuit (slow eye movement) keeps the image on the fovea. Congenital ocular motor apraxia (oculomotor apraxia, Cogan type) is a condition in which voluntary horizontal saccades are impaired and are associated with abnormal jerky head movements or thrusts, which enable fixation (4–6). Random saccades, vertical eye movements, and pursuit tend to be normal. Associated neurologic disorders include developmental delay and speech apraxia (6, 7).

Previous series have indicated a variety of anatomic abnormalities in children with congenital ocular motor apraxia, including dysgenesis of the corpus callosum, cerebellar hypoplasia, and posterior fossa tumor (6, 8–13). Children with Joubert syndrome—which is characterized clinically by developmental delay, episodic apnea, or tachypnea in the neonatal period, and radiographically by vermian agenesis or hypoplasia—may also have some features of congenital ocular motor apraxia (14). Children in whom magnetic resonance (MR) images show partial vermian agenesis without posterior fossa cyst may also exhibit ocular motor abnormalities, including ocular motor apraxia (15).

The purpose of this review was to determine the frequency of imaging abnormalities of the

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TABLE 1: CT and MR findings in 10 children with partial congenital ocular motor apraxia

Case	Age at CT, mo	Age at MR Imaging, mo	Thin-Section MR Imaging Performed*	Imaging Findings†
1	1	4, 51	Yes	Small inferior portion of vermis
2	6	150	Yes	Small inferior portion of vermis
3	9	14	Yes	Small inferior portion of vermis
4	12	87	Yes	Small inferior portion of vermis Small right cerebral hemisphere
5	13	49	...	Small vermis
6	29	...	...	Normal
7	49	...	...	Small vermis
8	103	...	...	Disorganized cerebellum
9	...	10	Yes	Normal
10	...	13, 28	Yes	Unclassified leukodystrophy

* Thin-section MR imaging comprised volumetric T1-weighted gradient-echo sequences with a 25.6-cm field of view, a 256 × 256 matrix, and 2-mm-thick sections.

† Vermian lesions in patients who had thin-section MR imaging are classified according to inferior or superior vermian involvement. Distinction between superior and inferior involvement was not considered accurate in patients who had only CT or conventional MR imaging.

brain in children with congenital ocular motor apraxia. We hypothesized that we would find a high frequency of abnormalities of the cerebellar vermis.

Materials and Methods

Thirty-two children, including two siblings, with the clinical diagnosis of congenital ocular motor apraxia were identified from the records of the visual impairment program of a tertiary care children's hospital. All the children were in the care of, or had been examined by, one pediatric neurologist with special interest in visual impairment. None of the children had breathing abnormalities suggestive of Joubert syndrome. All subjects had normal visual acuity. Our study group comprised the 19 children who had undergone brain imaging studies (12 boys and seven girls).

Congenital ocular motor apraxia was diagnosed when subjects showed signs of apraxic voluntary horizontal saccades, an absent or defective quick phase of optokinetic nystagmus, blinking prior to voluntary saccades, and characteristic head thrusting. All affected children also had truncal ataxia and hypotonia.

Expanded congenital ocular motor apraxia (7) was diagnosed when, in addition to the above findings, there were significant feeding difficulties or speech apraxia, or more severe generalized motor problems. On the basis of these criteria, nine children were categorized as having expanded congenital ocular motor apraxia. The other 10 children with typical congenital ocular motor apraxia were classified as having *partial* congenital ocular motor apraxia.

During a 13-year period, 15 of 19 children with congenital ocular motor apraxia were examined by computed tomography (CT), and 13 were examined by MR imaging. The median age at the first CT study was 12 months (range, 1 to 103 months); the median age at the first MR imaging study was 49 months (range, 4 to 175 months).

Axial CT scans were obtained 20° to the canthomeatal line using a 5- or 10-mm section thickness. MR studies were performed at 1.5 T. MR examinations included T1- and T2-weighted sequences in sagittal and axial planes. Later MR examinations included thin-section sagittal and/or coronal 2-mm-thick volumetric T1-weighted sequences and coronal fast spin-echo T2-weighted acquisitions. Inversion-recovery images were obtained in some cases. Contrast material was not administered.

All CT and MR studies were reviewed independently by two pediatric neuroradiologists who were blinded to the findings of clinical review other than the diagnosis of congenital ocular motor apraxia. Final diagnosis in each case was reached by consensus.

Results

Tables 1 and 2 list the patient information and imaging findings in the study group. Brain imaging studies were normal in five of the 19 subjects. Twelve of 19 children were found to have cerebellar abnormalities. The most distinctive finding in our patients was an abnormally small cerebellar vermis, which was seen in 10 cases (53%) on axial CT and MR studies (Fig 1A and B) and confirmed on sagittal or coronal MR images (Figs 1C and 2A and B).

Among the 10 patients with a small vermis, seven had thin-section sagittal or coronal MR sections through the posterior fossa. The inferior portion of the vermis was small or absent in all seven, while the superior portion was small and irregular in two of the seven (Fig 3). On

TABLE 2: CT and MR findings in nine children with expanded congenital ocular motor apraxia

Case	Age at CT, mo	Age at MR Imaging, mo	Thin-Section MR Imaging Performed*	Imaging Findings†
11	3	149	Yes	Small inferior and superior vermis
12	4	· · ·	· · ·	Small vermis
13	6	· · ·	· · ·	Partial agenesis of corpus callosum
14	10	91	Yes	Small inferior and superior vermis Pachygyria of right cerebral hemisphere
15	17	45	Yes	Normal
16	21	· · ·	· · ·	Hypoplasia of left cerebellar hemisphere Heterotopic gray matter in left lateral ventricle
17	39	175	Yes	Normal
18	· · ·	42	Yes	Small inferior portion of vermis Periventricular leukomalacia
19	· · ·	60	· · ·	Normal

* Thin-section MR imaging comprised volumetric T1-weighted gradient-echo sequences with a 25.6-cm field of view, a 256 × 256 matrix, and 2-mm-thick sections.

† Vermian lesions in patients who had thin-section MR imaging are classified according to inferior or superior vermian involvement. Distinction between superior and inferior involvement was not considered accurate in patients who had only CT or conventional MR imaging.

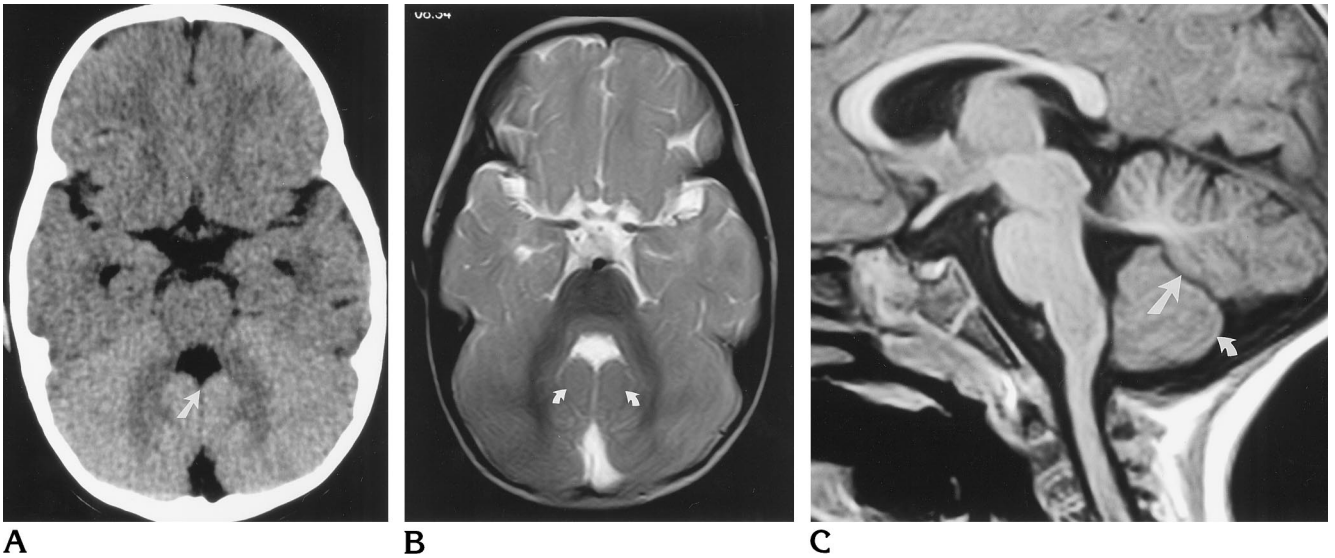

Fig 1. Case 3: Small inferior portion of the vermis in a 9-month-old boy with typical features of congenital ocular motor apraxia.

A, Axial 5-mm-thick CT scan shows midline posterior pointing of the roof of the fourth ventricle (*arrow*).

B, Axial 5-mm-thick T2-weighted fast spin-echo MR image (3555/80/2 [repetition time/effective echo time/excitations]) obtained 5 months later shows similar findings. The nodulus is absent and the cerebellar tonsils (*arrows*) are closely apposed.

C, Sagittal 2-mm-thick volumetric radio frequency (RF)–spoiled gradient-echo T1-weighted MR image (24/4.4/1; 30° flip angle) shows a normal-appearing superior portion of the vermis with absence of the anterior lobules of the inferior portion of the vermis (*straight arrow*). Thin sections allow separation of the abnormal inferior portion of the vermis from the adjacent cerebellar tonsils (*curved arrow*).

sagittal thin-section MR images of children with a small vermis, the superior cerebellar peduncles were elevated or horizontal in three of seven cases (Figs 1C and 2A), while on axial images the superior cerebellar peduncles appeared elongated in four cases (Figs 2C and 3B).

A small cerebellar vermis was found in six of 10 children with partial congenital ocular motor apraxia, and in five cases this was the only abnormality. A small cerebellar vermis was found in four of nine subjects with expanded congenital ocular motor apraxia, and was the only abnormality in two. Among the seven subjects with a small vermis in whom thin-section MR images were obtained, none of the four children with partial congenital ocular motor apraxia had superior vermian involvement,

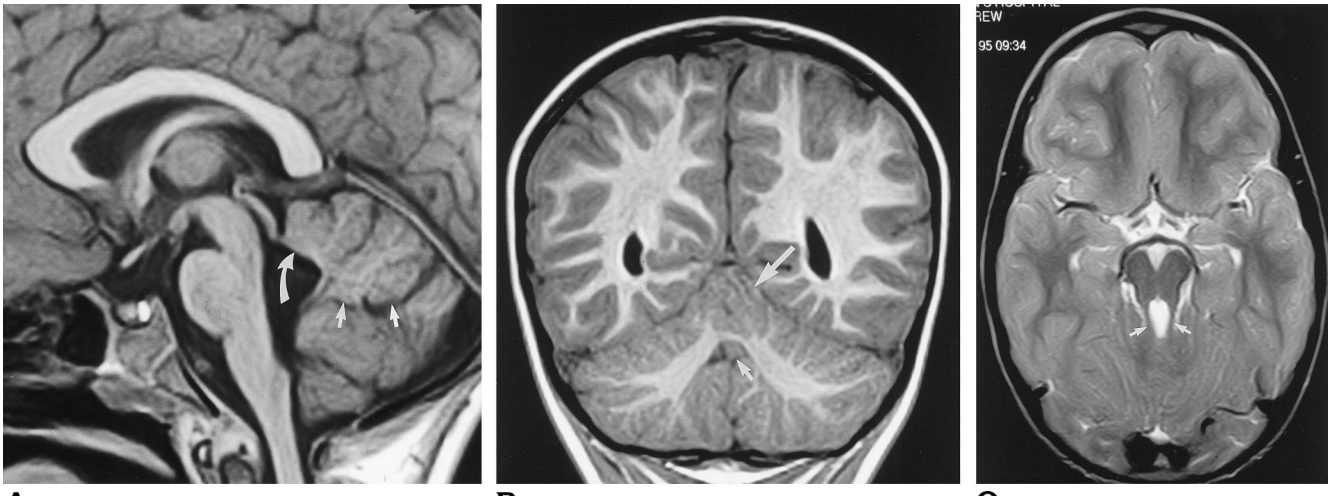

Fig 2. Case 1: Small inferior portion of the vermis in a 4-year-old boy with partial congenital ocular motor apraxia who presented initially with head titubation and nystagmus at 5 days of age. CT scan at 1 month (not shown) was interpreted as normal.

A, Sagittal 2-mm-thick volumetric thin-section volumetric RF-spoiled gradient-echo T1-weighted MR image (24/4.4/1; 30° flip angle) shows the inferior portion of the vermis (*straight arrows*) to be markedly smaller than the superior portion. The apposing cerebellar hemispheres and tonsils are seen below the inferior portion of the vermis. There is mild ballooning of the roof of the fourth ventricle (*curved arrow*).

B, Coronal 2-mm-thick volumetric RF-spoiled gradient-echo T1-weighted MR image (22/4.4/1; 30° flip angle) shows the marked disproportion of superior (*large arrow*) and inferior (*small arrow*) portions of the vermis. The superior portion appears symmetric (compare Fig 3).

C, Axial 5-mm-thick T2-weighted fast spin-echo MR image (3555/80/2) shows mild elongation of the superior cerebellar peduncles (*arrows*).

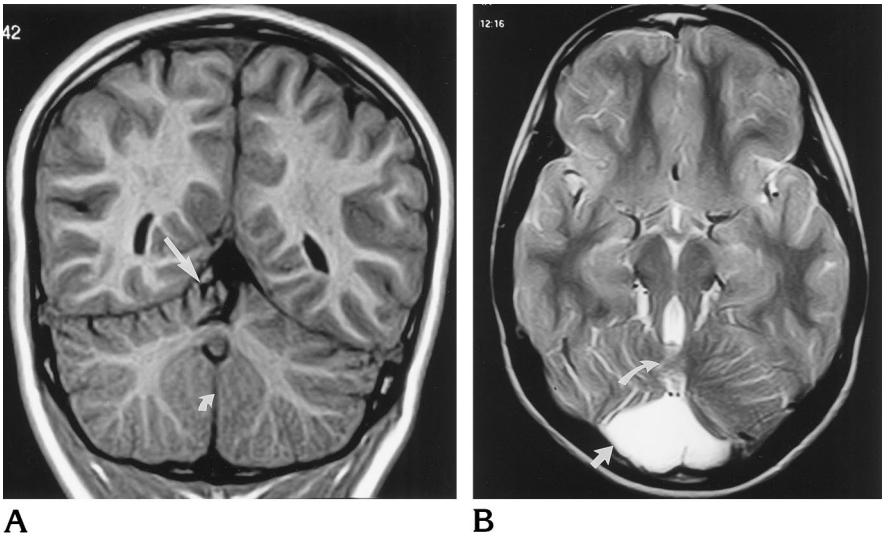

Fig 3. Case 11: Small vermis in a 12-year-old boy with expanded congenital ocular motor apraxia who was initially thought to have visual impairment. He had hypotonia and developmental delay. In retrospect, a CT scan at the age of 3 months (not shown) revealed midline posterior pointing of the fourth ventricle.

A, Coronal 2-mm-thick volumetric RF-spoiled gradient-echo T1-weighted MR image (22/4.4/1; 30° flip angle) shows both the superior and inferior portions of the vermis to be small. The superior portion is irregular (*straight arrow*), suggesting atrophy or a destructive pathogenesis rather than hypoplasia. Note the exaggerated vertical cleft (*curved arrow*) between the cerebellar hemispheres caused by the small inferior portion of the vermis.

B, Axial 5-mm-thick T2-weighted fast spin-echo MR image (3555/80/2) shows elongation of the superior cerebellar peduncles. There is a prominent cisterna magna (*straight arrow*). The small superior portion of the vermis is noted (*curved arrow*).

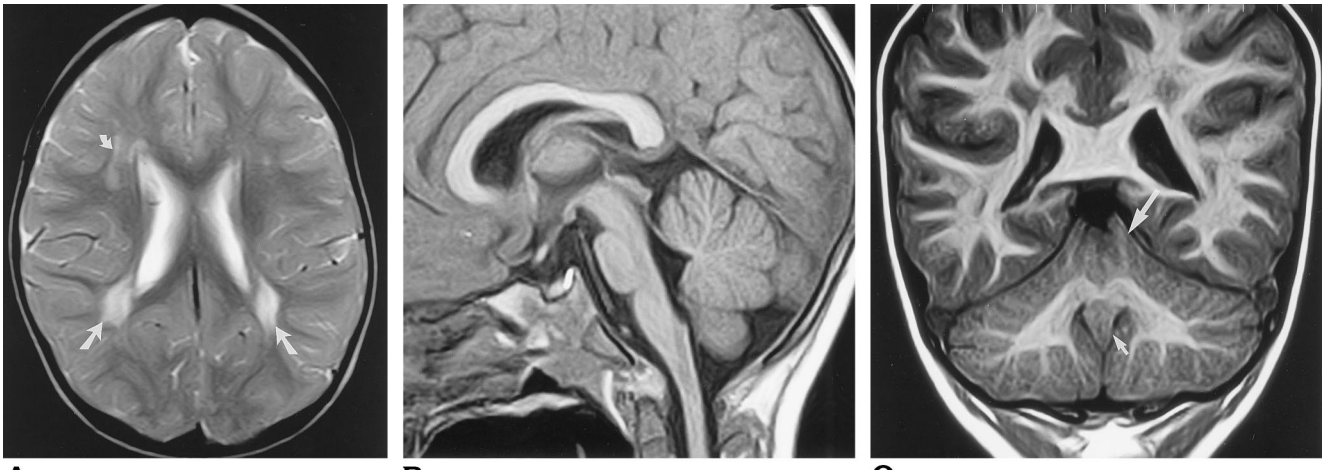

Fig 4. Case 10: 2-year-old boy with partial congenital ocular motor apraxia, abnormal white matter, but a normal cerebellum.

A, Axial fast spin-echo T2-weighted MR image (3555/80/2) shows symmetric hyperintensity in the periventricular white matter (*arrows*). The child was not premature and there was no white matter volume loss on T1-weighted sequences. A presumptive diagnosis of leukodystrophy was made, but no biochemical abnormality was identified.

B, Sagittal 2-mm-thick volumetric T1-weighted MR image shows normal size of the vermis with a normal configuration of the fourth ventricle. Compare with the abnormal inferior portion of the vermis shown in Figures 1C and 2A.

C, Coronal 2-mm-thick volumetric T1-weighted MR image again shows abnormal signal in the periventricular white matter. Note also normal separation of the cerebellar tonsils and medial cerebellar hemispheres by the inferior portion of the vermis (*small arrow*); the superior portion is symmetric and of normal size (*large arrow*).

while two of three subjects with expanded congenital ocular motor apraxia had superior vermian abnormality.

Two subjects had other cerebellar abnormalities. One of these children had nonspecific disorganization of both the cerebellar vermis and hemispheres on CT studies, but further detailed study by MR imaging was not performed. The other child had a normal-appearing vermis with hypoplasia of the left cerebellar hemisphere.

Of the 10 children with an abnormally small vermis, three had associated supratentorial lesions (see Tables). One of the two children with other cerebellar abnormalities also had a supratentorial lesion. Two children had supratentorial lesions with normal findings in the posterior fossa; these were partial agenesis of the corpus callosum in one and unclassified leukodystrophy in the other (Fig 4).

Discussion

The neurologic control of horizontal eye saccades is complex and incompletely understood. Interneurons connect the ocular motor nuclei via the medial longitudinal fasciculus and coordinate eye movements at a peripheral anatomic level. The supranuclear center for horizontal saccades is the paramedian pontine reticular formation. Input to this structure originates in the frontal eye fields, the vestibular nuclei, the superior colliculi, the cerebellum, the perihypoglossal nuclei, and the rostral interstitial nucleus of the median longitudinal fasciculus. In addition, horizontal eye movements can be affected by the posterior parietal lobe, the supplementary motor cortex, the thalamus, and the basal ganglia (16).

Congenital ocular motor apraxia is diagnosed when a typical constellation of eye and head movement abnormalities develops during the first few months of life (4, 6). Affected infants are visually inattentive and may be thought to be blind. Signs of cerebellar dysfunction, such as ataxia and hypotonia, emerge in early childhood. While both autosomal recessive and dominant inheritance are reported, we note that only two of our subjects were related (5). We have not performed imaging studies in unaffected siblings.

Ocular motor apraxia tends to improve with time, but does not entirely disappear. The compensatory head thrusting diminishes in most patients but can be seen with tiredness or anxiety; in older children, blinking tends to replace head thrusting. It is our experience that the generalized difficulties of hypotonia, ataxia, and coordination improve with age, but learning problems frequently emerge during the early school years. Children with congenital ocular motor

apraxia often need multidisciplinary assistance, including physiotherapy, speech therapy, and special education, as normal eye movements are important for reading.

Rappaport et al (7) reported that eight of 10 children with congenital ocular motor apraxia had other difficulties of motor organization, most marked in oral motor planning. These authors found that while ocular motor apraxia improved with time, problems with speech production continued. For the purpose of this review, therefore, our subjects with congenital ocular motor apraxia were divided into two groups. Partial congenital ocular motor apraxia included the typical ocular motor apraxia, ataxia, and hypotonia; expanded congenital ocular motor apraxia included affected children who also had speech apraxia or more profound feeding or motor difficulties.

We found cerebellar lesions in 12 of 19 subjects. An abnormally small cerebellar vermis was the most common finding. Like other authors, we believe the small vermis is usually the result of hypoplasia or partial agenesis. While vermian hypoplasia may be diagnosed on both axial CT and MR studies (Fig 1), we found that the diagnosis had commonly not been made prospectively. Thin-section (2-mm) sagittal and coronal MR images (Figs 1C and 2A and B) in some of our subjects showed that vermian abnormality preferentially involved the inferior lobules VII to X (tuber, pyramis, uvula, and nodulus) (17). With axial CT or conventional 5-mm-thick sagittal MR imaging, we found that we could not confidently distinguish inferior from superior vermian abnormalities.

The superior portion of the vermis was found to be small in only two of seven subjects in whom thin-section MR images of the cerebellum were available. In these two, the superior portion of the vermis was irregular and asymmetric (Fig 3), and we speculate a destructive pathogenesis or atrophy rather than hypoplasia. However, in no case did we find a progressive lesion during follow-up of between 6 months and 12 years. One of our two patients who had a superior vermian abnormality also had unilateral cortical pachygyria but no evidence of cortical atrophy.

Vermian agenesis or hypoplasia has been described before in children with congenital ocular motor apraxia, but the frequency in our series appears greater than previously described (6, 9, 13). The most characteristic finding is midline posterior pointing of the fourth ventricle at the level of the cerebellar peduncles on axial images (Fig 1A and B). This is due to loss of the posterior impression made by the nodulus of the inferior portion of the vermis (Fig 1C). In coronal sections, there is superior extension of the usual inferior midline cleft between the cerebellar tonsils (Fig 3A). The superior cerebellar peduncles may be elongated (Figs 2C and 3B).

We found a tendency for an isolated abnormality of the inferior portion of the vermis to be more common in children with partial congenital ocular motor apraxia than in those with expanded congenital ocular motor apraxia, but the difference was not statistically significant. Both children shown to have superior vermian involvement had expanded congenital ocular motor apraxia. Multiple centers in the brain, including the cerebellar vermis, are known to be involved in the control of eye movements (2, 16). In adult patients with posterior vermian lesions, abnormal saccades may develop as a result of cerebellar infarction (3). Our observations provide additional evidence that the control of horizontal saccades involves the inferior portion of the vermis.

Vermian agenesis or hypoplasia is well described in children with Joubert syndrome, which may include ocular motor abnormalities. In contrast to our patients, children with Joubert syndrome are said to show more severe involvement of the superior portion of the vermis, with superior convexity of the fourth ventricle; the superior cerebellar peduncles appear to arise nearly at right angles from the brain stem (14). Mild elevation of the superior cerebellar peduncles was noted in three of our seven patients who had high-resolution sagittal MR imaging (Fig 2A), although in two of these the superior portion of the vermis was normal on coronal images. A small medulla and upper cervical cord are also described in children with Joubert syndrome, and were not seen in our patient group. Despite these differences, we do not believe it is possible to distinguish patients with congenital ocular motor apraxia from those with Joubert syndrome by imaging criteria alone. None of our patients had the respiratory symptoms or retinopathy of classical Joubert syndrome.

In a recent series of 11 children found at MR imaging to have vermian hypoplasia without posterior fossa cyst, five had ocular motor apraxia and all 11 had disorders of ocular

movement (15). The inferior portion of the vermis was more affected than the superior portion, but images in that report show more severe involvement of the superior portion of the vermis than in our cases.

Vermian hypoplasia or agenesis may be isolated or associated with other central nervous system or peripheral malformations (18). The most common of these is Dandy Walker complex, which consists of partial or complete vermian agenesis with a posterior fossa cyst. We did not find any patient with a large posterior fossa cyst, although one with a small vermis did have a large cisterna magna (Fig 3B). None of our patients had ventriculomegaly. None of the syndromes associated with vermian agenesis was identified, and unlike with children who have vermian agenesis on prenatal sonograms, there were no children with karyotypic abnormalities (19).

Miscellaneous supratentorial abnormalities were found in six of our 19 patients (Tables 1 and 2). These included lesions affecting both the gray and the white matter, and were seen both in isolation and also with associated posterior fossa abnormalities. No anatomic location was consistently affected. We found no difference in the frequency of supratentorial abnormalities between patients with partial and expanded congenital ocular motor apraxia. Cerebral abnormalities included developmental lesions, such as pachygyria and subependymal heterotopia, and destructive lesions, such as periventricular leukomalacia and leukodystrophy. Contrary to early reports, an abnormality of the corpus callosum was identified in just one patient with partial agenesis (10, 20). These findings reinforce the opinion that congenital ocular motor apraxia is a symptom or sign rather than a specific disease entity (12). The variety of lesions detected is in keeping with the large number of sites involved in eye movement control.

In this study we reviewed the imaging findings in children with typical congenital ocular motor apraxia. Three other children thought to have congenital ocular motor apraxia were initially included in our radiologic analysis; however, on clinical review performed without knowledge of the radiologic findings, these children were found to have atypical features, and were subsequently excluded from the series. Imaging findings in these three, determined prior to knowledge of the atypical clinical features, appear to be different. In two, there was atrophy of the whole vermis and cerebellar hemispheres, while the findings in the third child were considered normal.

CT and MR imaging techniques in this series were variable and were more detailed on the more recent studies. Furthermore, not all children had MR imaging, and our early studies were performed with a view to assessing the corpus callosum. It is possible, therefore, that some developmental anomalies of the cerebellar vermis may have gone undiagnosed. In the detailed MR assessment of the posterior fossa in children with congenital ocular motor apraxia, we would now recommend the following to enable more accurate delineation of the anatomy of the lobules of the vermis: 3-mm high-resolution sagittal or coronal fast spin-echo T2-weighted sequences, and sagittal or coronal volumetric T1-weighted sequences using a 1- or 2-mm section thickness.

In summary, we have presented the imaging findings in 19 children with congenital ocular motor apraxia. Like previous authors, we found this disorder to be a symptom associated with a number of different imaging findings. While there is no single unifying radiologic diagnosis, we did find a high prevalence of abnormally small cerebellar vermis. We found preferential abnormality of the inferior portion of the vermis, which we ascribe to hypoplasia or partial agenesis. Our observations support the view that the control of horizontal saccadic eye movements involves the inferior portion of the vermis.

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