Mutism in an Adult Following Hypertensive Cerebellar Hemorrhage: Nosological Discussion and Illustrative Case

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Mutism after cerebellar injury has been associated with tumors, hemorrhage, and surgery of midline cerebellar structures. Literature review identified 54 cases, primarily in children after surgical splitting of the inferior vermis. We present a 47-year-old who developed transient mutism after cerebellar hemorrhage. This represents the first report of transient mutism in an adult with neither tumor nor brainstem infarction and documents the importance of cerebellar structures for initiation and production of speech in adulthood. This case further differs from those previous because of the long mute period and the subsequent return of continued ataxic and dysarthric speech. © 1997 Academic Press

INTRODUCTION

The syndrome of cerebellar mutism following tumor, surgery, or hemorrhage consists of a brief interval of normal speech, followed by weeks or months of mutism. The returning speech may be normal or very dysarthric (Rekate, 1985; Yonemasu, 1985; van Dongen, 1994). Patients are all alert with severe cerebellar ataxia, but have no long tract signs, cranial nerve palsies, nystagmus, or other signs of brainstem involvement. The inferior vermis, and/or the dentate nuclei and their related pathways are putative sites of this speech deficit. Magnetic resonance imaging does not reveal brainstem involvement (e.g., infarction, hemorrhage, demyelination, cysts, etc.). The syndrome is important to recognize because of both its severity and transient nature. The pathogenesis remains unclear. Our review of the literature identified 54 cases of cerebellar mutism, occurring primarily in children after surgical splitting of the inferior vermis.

We present a case of transient mutism in an adult who suffered a cerebellar

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hemorrhage. To our knowledge, this case represents the first report in an adult without a cerebellar tumor or brainstem abnormalities who had the onset of his mutism following a period of speech competence after his ictus. This case further differs from those previous because of the long mute period and the subsequent return of continued ataxic and dysarthric speech.

REPORT OF A CASE

While gardening, a 47-year-old right-handed man experienced abrupt onset of severe left occipital headache accompanied by garbled speech, diaphoresis, and left-sided weakness. Paramedics found him alert, following commands, and conversant; systolic blood pressure was 190 mm Hg. He developed hyperemesis and had endotracheal intubation. In the emergency room, his blood pressure was 200/118. On examination his pupils were equal, round, and symmetrically reactive to light; he visually fixed and tracked; extraocular movements were intact without nystagmus; corneal reflexes and face were symmetric; a weak gag reflex was present; extremities were of normal tone, bulk, and strength; sensory examination and deep tendon reflexes were normal and symmetric; plantar responses were flexor bilaterally. He had gait ataxia and dysarthria.

Admission computed tomography (CT) of the head showed a large superior vermian hemorrhage, extending symmetrically and anteriorly to the brachium pontis (Figs. 1A and 1B). The effaced fourth ventricle contained a small volume of fresh blood; there was no subarachnoid blood. Mild enlargement of the lateral and third ventricles was present. Mild ponto-mesencephalic mass effect was present, but the prepontine, interpeduncular, and ambient cistems were visible. Neither intravenous contrast infusion CT scan nor cerebral angiography demonstrated an aneurysm or arteriovenous malformation.

In the operating room, a right frontal ventriculostomy released xanthochromic cerebrospinal fluid under high pressure. Immediately postoperatively, he became unresponsive with pinpoint pupils and decorticate posturing. Repeat CT scan showed increased midline posterior fossa blood, extending bilaterally into the region of the brachium pontis and superior vermian cistern. The brainstem appeared under compression. He returned to the operating room, where a suboccipital craniectomy permitted evacuation of a large clot. Midsagittal splitting of the vermis exposed the fourth ventricle. Pathological examination of the surgical specimen showed only clot and normal cerebellum.

Postoperative CT scan confirmed removal of a majority of the superior vermian clot, demonstrating only mild dilation of the lateral ventricles, without evidence of infarction or new hemorrhage (Fig. 1C).

Postoperatively, the patient was intermittently awake, occasionally following commands and remained intubated. After removal of the ventriculostomy on the tenth postoperative day, a contrast-enhanced CT did not demonstrate hydrocephalus, abscess, or infarction.

A waxing and waning level of consciousness, which slowly improved, marked his post-operative course. Two weeks post-operatively, magnetic resonance imaging demonstrated a $3 \times 5 \times 15$ mm area of old hemorrhage occupying most of the vermis and displacing the fourth ventricle anteriorly (Figs. 2A and 2B). Edema was evident in the mesial cerebellar hemispheres and middle cerebellar peduncles. There were no new infarcts.

After 3 weeks, he had slight left arm weakness and dyscoordination. He spontaneously opened his eyes and tracked, followed commands, and responded appropriately to "yes/no" questions with hand and head motions. He did not vocalize. Swallowing evaluation showed swift laryngeal elevation with reflexive swallowing after throat clearing. He continued to require naso-duodenal feeding.

Neurological examination showed minimally decreased lateral excursion in both eyes. Upgaze and pupillary examinations were normal. He had marked symmetric orofacial dyscoordination, affecting the purposeful use of his tongue and his ability to mouth words. He had a weak symmetric gag reflex. His tongue protruded in the midline. He was able to stand and take a couple of steps but only with two-person assistance. He had marked ataxia of all his limbs, with past-pointing, dysmetria, dysdiadochokinesia, dyscoordination and prominent dyssynergia. Check reflexes were poor in all limbs. Romberg's sign was present, and he had postural instability with poor maintenance of station. Fine finger movements were poor. Obvious paresis was absent. Deep tendon reflexes were symmetric and Babinski's sign was absent bilaterally.

Head CT scan that day did not reveal any evidence of hydrocephalus, midline shift, or mass effect. The midline vermis and cerebellar hemispheres, left more than right, demonstrated hypodense areas. The brainstem appeared normal.

He exhibited intermittent agitation. He responded appropriately to verbal questions, was able to perform multistep commands, and could communicate using his head and upper extremities. Except for occasional grunting, he was mute. He could sort and categorize objects. Severe ataxia limited his writing. The patient's reading comprehension appeared normal, responding by way of a communication board or head nodding. He accurately used a needs board, with a light pointer, and an eye gaze board for creating novel messages. He needed supervision to organize his directional eye gaze and exercise control over his ataxia and frustration to use these devices. He could stabilize a movement or posture when given time. He could use the eye gaze board to communicate 11 single word and phrase length messages over 15 min. He was able to follow commands. He was able to spell correctly three words in structured tables. He could perform convergent single-word language tasks. He correctly performed a name-the-capitol/state exercise with



FIG. 1. (A) Axial, unenhanced CT scan demonstrates a superior vermian hematoma extending to fill the vallecular cistern. (B) The hematoma extends superiorly to the quadrigeminal cistern. The prominent temporal horns suggest incipient hydrocephalus. (C) Post-operative axial CT scan demonstrates the evacuation of the hematoma.



FIG. 1—Continued



FIG. 1—Continued

the eye gaze board. He could sort and categorize objects. He did not receive forced-choice recognition memory testing. He remained completely non-speaking (mute).

Three months after his ictus, blood pressure was 138/100. The following were normal on examination: extraocular movements, hearing, tone, motor strength, tendon reflexes, primary modality sensation, and tongue strength. He remained mute. He could chew well, but his oropharyngeal coordination was too poor for swallowing, and he had no gag reflex. He received feedings by percutaneous gastrostomy. Ataxia precluded self-feeding. He had rare vocalizations, but no articulation of words. No mouthing of words was apparent. The patient communicated with head nods. Writing attempts were ataxic. He was able to calculate and identify objects. He was alert and oriented and not demented. Extensive psychological testing demonstrated no appreciable cognitive losses. He manifested prominent bouts of emotional lability. Incoordination of the tongue, face, and mouth was greater than that of trunk or extremities. He could not dress himself, apparently because of ataxia rather than dyspraxia. The patient could independently position himself in the bed. but, because of truncal ataxia, he would fall over if he swung his legs over the edge. He could stand for a shower but could only take a couple of steps with two-person assistance.

Barium cinematography revealed uncoordinated epiglottic inversion when swallowing liquids.

Follow-up of his risks for recurrent stroke revealed normal extracranial Doppler exam of his neck. Because of claustrophobia, the patient refused follow-up MRI.

Four and one-half months after his ictus, his mutism cleared as he uttered a comprehensible word. Four days prior, he began to mouth words without phonation. Over the ensuing 4 days, he was able to speak three names and two three-word phrases (''I love you;'' ''I can speak''). Over the following 4 days, he could speak 46 words with a prolonged ''ah'' sound for 5–7 sec. His ability to initiate speech and maintain phonation remained inconsistent. Effortful, forced exhalation with facial grimacing accompanied all speech attempts. All phonation was high in pitch and, at times, not coordinated with articulation. Articulatory errors appeared related to the manner of production with more errors occurring with sounds that were more complex in motoric production (e.g., sh, 1, r, s). A phonatory/articulatory ataxia, graded as a moderate/severe ataxic dysarthria, characterized his speech. He exhibited uncoordinated/inefficient breath support for speech. He continued to have marked bucco-lingual dyscoordination with dysarthria. He could reliably spell words vocally.

Ongoing cognitive assessment showed his ability to perform word associations, proverb interpretations, and nonverbal spatial reasoning. He could repeat complex sentences and recall facts from a story in a logical sequence.

After another month, he had barely comprehensible but very ataxic speech



FIG. 2. (A) Sagittal midline T1-weighted MRI study reveals the hyperintense signal of the residual hematoma involving the superior and midline vermis. Minimal mass effect is evident. Note the radiographically normal brainstem. (B) Axial T1-weighted MRI study demonstrates the residual hyperintense signal of the vermian hematoma. The fourth ventricle is not displaced.



FIG. 2—Continued

and was able to swallow solids. One year after his ictus, he could propel a manual wheelchair with his feet. He participated in conversation as a responder, rather than initiator, and kept his verbal responses (sometimes supplemented by gestures) brief. He would become emotional and tearful during speaking. His speech was characterized by great effort, ataxic dysarthria, and strained voice, with pitch often higher than normal. Intelligibility was approximately 75% with known content. He used spelling to remedy commu-

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| Test | Score (percentage correct) |
|---------------------|-------------------------------|
| Functional reading | 90 |
| Paragraph-picture | 100 |
| Factual-inferential | 100 |

 TABLE 1

 Scores of Patient on the Reading Comprehension

 Battery for Aphasia (LaPointe, 1979)^a

^{*a*}At 1 year post-ictus. Patient was no longer mute at the time.

nication breakdowns. He independently used an alphabet board to indicate the first letter of misunderstood words.

The patient followed complex spoken and written commands. He underwent formal language testing (Table 1). He self-corrected his speech errors, named without difficulty, and described object accurately. He demonstrated moderately decreased word fluency on a task requiring generation of members of a category. The patient's printing was marginally legible at the simple sentence level secondary to ataxia. He profited from the use of a typewriter to stabilize his arms, yet ataxia resulted in misstrikes of adjacent keys. Spelling, grammar, and punctuation were all intact. He did not require cues to operate the typewriter.

On immediate auditory memory tasks, the patient repeated five sentences of increasing complexity accurately and recalled in detail the events of two narrative paragraphs. He could complete two- and three-dimensional block designs. Visual attention span was consistent at five units, which is normal. On ideational tasks, he mentally shifted on a forward/backwards spelling task and completed six of six simple calculations correctly. He performed pictorial associations of increasing abstraction.

Over the 39 months since his initial ictus, he has made slow, steady improvement, but he still has marked ataxia of his trunk, extremities, and speech.

DISCUSSION

Our patient presents a picture of prolonged mutism without cranial nerve findings, and severe ataxia related to a cerebellar hemorrhage. Mutism followed a latent period after surgery that included vermian splitting. His speech has not returned to its premorbid normal pattern, remaining ataxic over longterm follow-up. He has also had persistent incoordination of his tongue and deglutition. The exact trigger is unclear. This case did not involve a tumor as have most of the other cases of cerebellar mutism.

Disorders associated with mutism. Psychiatric disorders account for the vast majority of mutism cases without brainstem signs (Benson, 1979). Organic causes usually have long tract signs and/or alterations of conscious-

ness. Seizures, which are extra-cerebellar in origin, may also cause speech arrest. Six sites or syndromes render mutism: Broca's aphasia (usually transient), the dominant hemisphere's supplementary motor area, the mesence-phalic reticular formation (associated with akinetic mutism), pseudobulbar palsy (secondary to diffuse bilateral cerebral hemispheric dysfunction), bilateral thalamotomy in Parkinsonism [Benson, 1979], and bilateral laryngeal paralysis.

The dominant hemisphere supplementary motor area syndrome includes buccomotor dyspraxia among the constellation of findings (Rostomily, 1991). Bilateral above the pons lesions can cause so-called pseudobulbar palsy, sometimes associated with mutism. This usually presents with upper motor neuron abnormalities and a flat, but labile, affect.

There are described speech abnormalities after posterior fossa injuries. Review of 40 patients with missile injuries to the cerebellum demonstrated that unilateral hemispheric lesions caused "slow, drawing, and monotonous" speech that was "staccato and scanning." Vermian involvement further distributed articulation and phonation. Bilateral hemispheric injury created speech that was nearly unintelligible (Holmes, 1917). The more recent reports on cerebellar mutism have described a brief interval of normal speech after cerebellar tumor surgery in children. The mute period has lasted 3 or more months followed by a severe derangement more characteristic of "cerebellar speech." Reversion to normal patterns ensued. The patients all were alert with "severe cerebellar ataxia," but "no remarkable brainstem signs." They had preserved phonation, intact comprehension, and could communicate through gestures (Yonemasu, 1985).

In past attempts to control spasticity and dyskinesias in cerebral palsy patients, bilateral stereotactic dentate nuclei ablation, sparing the lateral hemispheres, caused transient mutism (Fraioli, 1975; Guidetti, 1977). Speech returned over 1 to 3 months.

Central pontine myelinolysis may also involve extrapontine structures, such as the basal ganglia and the cerebellum. Mutism occurs in this syndrome, which may also include pseudobulbar palsy and restless agitation, besides the typical quadriplegia (Adams, 1959; Kepes, 1965). Disturbances of the cranial nerves originating in the pons help identify this form of mutism.

In the first description of akinetic mutism (caused by an epidermoid cyst of the third ventricle) the only utterances were monosyllabic whisperings (Cairns, 1941). Other cases of akinetic mutism occurred following surgery for tumors of the thalamus or hypothalamus, basal ganglia, midbrain, anterior cingulate gyrus, or third or fourth ventricles. These cases usually involved splitting the vermis at the level of the pyramis (Daly, 1958; Lavy, 1959; Skultety, 1968), leading to a warning to avoid the dentate nuclei when splitting the vermis (Dandy, 1966). Experimentally, involvement of the caudal ventromedial mesencephalic tegmentum, inferior quadrigeminal brachia and periaqueductal gray matter appears to cause this clinical picture (Skultety, 1968). Occlusion of the mesencephalic artery can also lead to akinetic mutism (Segarra, 1970).

The literature contains several cases of mutism of cerebellar origin, with the first reports appearing in the mid-1980s (Wisoff, 1984; Rekate, 1985; Yonemasu, 1985). Cerebellar mutism has been associated with tumors, hemorrhagic lesions, and surgery involving midline cerebellar structures primarily in children (mean age 12.3 years, median age 8 years; Table 2). The mute patient must be alert and show normal cognitive function. He must be able to comprehend, write, and read. Mutism is more than simply oral apraxia. This clinical syndrome is unusual with cerebellar disease. To implicate the cerebellum as the cause of this problem requires the absence of cranial nerve abnormalities, brainstem signs, dementia, or aphasia.

Review of 162 patients with nondegenerative cerebellar disease (e.g., hemorrhage, infection, tumor) provides evidence implicating the left anterior paravermian region in those with dysarthria (22 of 31 patients with left-sided cerebellar injury). Twenty-six of the 162 had primary vermian involvement, but only 3 of these patients had speech dysfunction, a finding similar to that of Gilman (1981). An autopsy series showed 70% prevalence of dysarthria in patients with left cerebellar hemispheric infarction (Sypert, 1975).

In 1985, a report appeared of six children with mutism and new cerebellar lesions, involving the vermis, hemispheres and deep nuclei (Rekate, 1985). There are a few earlier suggestions of this syndrome after childhood medul-loblastoma surgery (Hirsch, 1979; Wisoff, 1984; Yonemasu, 1985). Subsequently, other authors reported similar cases (Table 2). These patients predominantly have midline pathology. Although, not all case reports describe the surgical procedure, it appears that in roughly a third of cases there was surgical incision of the inferior vermis. Patients reportedly all spoke during the immediate postoperative period, before becoming mute 18–72 hr after operation. None had phonatory organ disturbance (Ferrante, 1990), but nearly 30% were unable to eat, even without lower cranial nerve palsies.

Our literature search located 54 reported cases of mutism associated with cerebellar lesions, almost all in children. Recent reviews found mutism complicating 6 of 105 (5.7%) (Cochrane, 1994) and 9 of 110 (8.2%) (Dailey, 1995) of aggressive pediatric posterior fossa tumor surgeries. The only adults previously described with mutism had juvenile tumors (two 20-year-olds and one 45-year-old with medulloblastomas and a 22-year-old with a pineal-oblastoma) requiring midline approaches (Salvati, 1991; D'Avanzo, 1993), or brainstem ischemia (a 48-year-old) (D'Avanzo, 1993).

Cognition and cerebellar mutism. Several authors describe cerebellar hypoplasia in various states of cognitive impairment, such as neocerebellar posterior vermian hypoplasia in autistic people (Courchesne, 1988), and similarly in Fragile X patients (Reiss, 1988) and learning-disabled children (Valk, 1988). Children with midline cerebellar hypoplasia have limited vocabularies and significant speech delays (Sarnat, 1980). These situations raise

the question of the role that the posterior vermis might play in acquiring speech skills.

In olivopontocerebellar atrophy (OPCA), positron emission tomographic (PET) studies demonstrate a correlation between the ataxia of speech and hypometabolism in the cerebellar vermis and hemispheres, and the brainstem, but not in other brain structures, such as the parietal cortex. The association between this and the degree of anatomical cerebellar change is weaker (Gilman, 1993). Neuropsychiatric evaluation of OPCA patients suggested some occult intellectual deficits, including verbal memory and intelligence as well (Kish, 1988). Motor learning seems likewise impaired in these patients (Sanes, 1990).

Motor and cognitive functions seem to have distinct somatotopic representations in the cerebellar cortex. Other PET studies have defined bilateral posterior vermian and inferolateral hemispheric activation with single word generation and the cognitive imagery before speech initiation (Petersen, 1988).

In normal adults, during word association, blood flow increases in the superior anterior lobule of the cerebellar cortex just lateral to areas where similar changes occur with eye or finger movements. The lateral inferior right cerebellar cortex remains similarly active when subtracting motor activity (tested separately) from these blood flow studies. This suggests a pure cognitive function to this area (Petersen, 1988). Thus the cerebellum appears involved in the functions of speech coordination, initiation, planning, and timing.

Anatomy and postulated mechanisms. The areas believed responsible for the speech disturbances of autistic patients and those with other nondegenerative cerebellar diseases are Larsell's lobules H VI and VII (Gilman, 1981; Courchesne, 1988). Auditory and visual stimulation also make these posterior vermian areas active (Snider, 1944; Altman, 1976). Spinocerebellar tracts pass by way of these areas (the pyramis, tuber, and uvula) which also receive sensory input from the head. Spindle afferents from the tongue and the other muscles of speech production also project there, by way of the mesencephalic nucleus of the trigeminal nerve. These areas also receive input from cerebral sensorimotor areas (Allen, 1974) (Figs. 3A and 3B). The adjacent simplex lobule contributes to laryngeal motor outflow (Larson, 1978). Left cerebellar hemispheric lesions may thus not be clearly responsible for mutism but may be so for dysarthria (since they coordinate the sensory input for the outflow of monitored speech patterns).

Cerebral dysfunction along the above-mentioned cerebellar sensory input pathways thus could explain mutism; however, our patient had no suggestion of such abnormality either on examination or imaging studies. Pontine nuclei, the ventrolateral thalamus, and cerebral cortex (supplementary motor and other areas) connect with the dentate (Carpenter, 1991). The intracerebellar connections between the dentate nuclei and the vermian and paravermian regions parallel the pathways between the cortex and the nuclei interpositi

| | | Reporte | TABLE 2 ed Cases of Cerebellar Mutism | | |
|-------------|--------------------------------|------------------|--|-----------------|--------------------|
| Case No. | 1st author/year | Age (yr), sex | Lesion location and histology | Latency (hr) | Mutism duration |
| 1* | Wisoff, 1984 | 6 M | Midline, IV vent | 48 | 2 wk |
| ř | | | Malignant astrocytoma | ç | . j |
| * * | W ISOII, 1984 W/isoff 1084 | 12 M 3 5 M | Vermis, L I V medulloblastoma Vermis IV medulloblastoma | 71 | 2.2 WK 10 dave |
| . * * | W ISULL, 1704 W/:ff 1004 | | Venues, 1V incouncertained | 04 0 | 10 uays |
| t ν. | w isolit, 1964 Rekate: 1985 | 1 / M 8 F | venuus, L I v venu asu ocytoma Vermian medullohlastoma | 40 84 84 | 3 months |
| 9 | Rekate, 1985 | 6 M | Vermis, L hemisphere | 72 | 3 months |
| | × | | Cystic astrocytoma | | |
| 7 | Rekate, 1985 | 2^{-} | -, Ependymoma | | 2 months |
| 8 | Rekate, 1985 | 10 M | -, Medulloblastoma | | 2 months |
| 6 | Rekate, 1985 | -6 | -, Medulloblastoma | | 3 months |
| 10 | Rekate, 1985 | 11- | Medulloblastoma | | 3 wk |
| 11 | Yonemasu, 1985 | | Bilateral ependymoma | 18 - 72 | 4-12 wk |
| 12 | Yonemasu, 1985 | | Bilateral ependymoma | 18 - 72 | 3 wk |
| 13 | Yonemasu, 1985 | | Bilateral medulloblastoma | 18 - 72 | 4-12 wk |
| 14 | Yonemasu, 1985 | | Bilateral medulloblastoma | 18 - 72 | 4-12 wk |
| 15 | Volcan, 1986 | 8 F | IV vent medulloblastoma | 24 | 2 wk |
| 16 | Humphreys, 1989 | 7 M | Vermis, R hemisphere | 24 | 16 wk |
| | | | Medulloblastoma | | |
| 17 | Humphreys, 1989 | 3 M | IV vent medulloblastoma | | 7 wk |
| 18 | Humphreys, 1989 | 7 M | IV vent medulloblastoma | | 10 wk |
| 19^{*} | Humphreys, 1989 | 4.5 M | Vermis, IV vent | 72 | 7 wk |
| | | | Fibrillary astrocytoma | | |
| 20^{*} | Humphreys, 1989 | 10 F | Vermis, IV vent ependymoma | 24 | 10 wk |
| 21 | Ammirati, 1989 | 14 F | Vermis, IV vent | 48 | 6 wk |
| | | | Pilocytic astrocytoma | | |
| 22 | Ferrante, 1990 | 9 F | Vermis, MCH, IV vent | 48 | 4 days |
| | | | Pilocytic astrocytoma | | |
| 23 | Ferrante, 1990 | 5.5 F | Vermis, MCH, IV vent | 48 | 8 days |
| 24 | Ferrante 1990 | θM | Pilocytic astrocytoma Vermis IV vent R hemisphere | 36 | 8 davs |
| t 1 | | | Pilocytic astrocytoma | > | o and o |

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| Dietze, 1990 | / M | vermis, MCH, IV vent Medulloblastoma | | 0-12 WK |
|----------------------------|--------|--|--------------|----------------|
| Dietze, 1990 | 15 F | Vermian AVM | Ι | 3 months |
| Salvati, 1991 | 20 M | Vermis, IV medulloblastoma | 46 | 4 wk |
| Nagatani, 1991 | 4 F | Vermian medulloblastoma | 24 | 78 days |
| Herb, 1992 | 9 M | Vermis, IV medulloblastoma | 12 | 10 wk |
| Catsman-Berrevoets, 1992 | 6 M | Vermis, IV medulloblastoma | 24 | 6 wk |
| Catsman-Berrevoets, 1992 | 8 F | Vermian medulloblastoma | 48 | 6 wk |
| Catsman-Berrevoets, 1992 | 8 M | Vermis, IV medulloblastoma | 24 | 8 wk |
| D'Avanzo, 1993 | 20 F | IV vent medulloblastoma | 48-72 | 5 months |
| D'Avanzo, 1993 | 45 M | Vermis, R hemisphere, IV vent Medulloblastoma | 68 | 3 months |
| D'Avanzo, 1993 | 22 M | R mesencephalon pinealoblastoma | 72 | 4–6 months |
| D'Avanzo, 1993 | 48 M | R ponto-mesencephalic ischemia | 72 | 16 wk |
| Cakir, 1994 | 61 M | R hemisphere metastasis | | 4 days |
| van Dongen, 1994 | 10 M | R hemisphere, peduncle, IV vent | | 2 days |
| | | Ependymoma | | |
| van Dongen, 1994 | 4 F | Ventral posterior fossa | | |
| van Dongen 1994 | βM | Meningtoma IV vent medullohlastoma | 74 | 5 wk |
| van Dongen. 1994 | 8 F | Vermian medulloblastoma | 48 | 5 wk |
| van Dongen, 1994 | 8 M | Vermis, IV vent medulloblastoma | 24 | 2 wk |
| van Dongen, 1994 | 5 M | Vermian medulloblastoma | <24 | 8 wk |
| van Dongen, 1994 | 4 M | IV vent, R medulla ependymoma | 24 | 3 wk |
| Silveri, 1994 | 67 M | R hemisphere infarction | with ictus | 'few hours'' |
| Dailey, 1995 | 4 M | -, Ependymoma | 24 | 2 wk |
| Dailey, 1995 | 12 F | -, Medulloblastoma | immediate | 1 wk |
| Dailey, 1995 | 9 F | -, Medulloblastoma | 12 | 4 wk |
| Dailey, 1995 | 6.75 F | Medulloblastoma | 12 | 12 wk |
| Dailey, 1995 | 3 M | Medulloblastoma | 24 | 1.5 wk |
| Dailey, 1995 | 6 F | Astrocytoma | 24 | 6 wk |
| Dailey, 1995 | 2.5 M | -, Medulloblastoma | 48 | 3 wk |
| Dailey, 1995 | 20 F | -, Astrocytoma | 12 | 8 wk |
| Dailey, 1995 | 10 F | -, Medulloblastoma | 09 | 3 wk |
| Coplin, 1997. Present case | 47 M | Vermian hemorrhage | - (see text) | 4.5 months |
| | | Average | 35.6–41.2 hr | 46.6–51.6 days |

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L, left; R, Right.



FIG. 3. Schematic representations of cerebellar bidirectional connections with the frontal cerebral cortex (A) and other brain areas (B) purported to be involved in speech production.

and fastigial nuclei (Tolbert, 1978). The pars intermedia receives from the spino- and cuneo-cerebellar tracts as well as the cerebrum, seemingly calibrating motor output (Allen, 1974; Ghez, 1991).

The role of the cerebellum in coordinating sensory input and executable motor outflow, thus planning the execution of motor speech, may be at play in causing cerebellar mutism. Surgical splitting of the cerebellum breaches the corpus medullarae of the inferior deep cerebellar white matter. The corpus medullarae is part of a more-complex system connecting not only the adjacent ipsilateral lobules but also the contralateral infero-lateral areas of the two hemispheres (Eager, 1963; Carpenter, 1991; Ghez, 1991). Feline studies further suggest such collateral cerebellar communication (Rosina, 1984) to the neocerebellum and the postero-central vermis (which includes the pars intermedia and Larsell's, lobules V and VI) by way of pontine nuclei. The supplementary motor and premotor areas also give to these pontine nuclei. This would explain intra- and trans-cerebellar communication in coordination with cortical motor planning and outflow. Additionally, that some zones are phylogenetically more recent could help explain their involvement in not only motor aspects of speech but also the cognitive aspects of language (Leiner, 1993). Such "commissural" disruption may interrupt the bilateral loop involved in the anatomy of the buccofacial dyspraxia seen after bilateral

injury of the cerebral frontal association cortices (Groswassa, 1988; Mao, 1989).

Work elucidating the role of the cerebellum in speech continues. It appears to serve to adjust the tempo and force of the finely coordinated muscular movements involved in speech production (Brown, 1949). Dysarthria, monotony, scanning, hesitancy and explosive speech most commonly mark cerebellar speech disorders (Flourens, 1823; Charcot, 1877; Holmes, 1917; Holmes, 1922; Zentay, 1937).

Some evidence points to the superior vermis and/or cerebellar hemispheres as the site(s) of these prevailing cerebellar speech deficits (Mills, 1914; Holmes, 1917; Holmes, 1922; Lechtenberg, 1993). Vermian or paravermian lesions alone may not be sufficient to cause mutism. Dysarthria may occur with lone cerebellar hemispheric lesions, but patients with vermian involvement seem to have greater speech difficulty (Holmes, 1917; Holmes, 1922). Paravermian and lateral hemispheric cerebellar tumors are most likely to cause dysarthria, while vermian damage is an uncommon cause of speech disorders (Amici, 1976). Some authors implicate the superior left hemisphere more than other areas (Lechtenberg, 1978; Ferrante, 1990). The left superior paravermian region receives inputs from cranial nerves VIII and X, as determined by electrical stimulation studies. The midportion of the left superior paravermis appears to coordinate sensorimotor and cortical inputs, particularly auditory and visual ones (Dow, 1939; Lam, 1952; Kent, 1976). One suggestion is that the posteromedial paleodentate, modulating preprogrammed motor speech commands from the cerebrum, mediated by interconnections with the vermis and paravermis, might provide a memory of the learned movements for speech. Injury to this circuit might abolish motor speech (Eccles, 1969; Ito, 1970; Dietze, 1990-1991).

"Cerebellar speech" is dysarthric and dysrhythmic, with inappropriate emphases. Scanning speech is an example of this, in which there is deliberate correction of the motor speech program. This disturbed coordination disturbs fluency. Comprehension and language functions are undisturbed. Scanning speech, as seen in multiple sclerosis, or ethanol intoxication, appears most related to left-sided paravermian cortex (Lechtenberg, 1978). An apparent inconsistency is that the left cerebellum interacts with the right thalamus and right cerebrum, whereas cerebral language functions predominate on the left (Lechtenberg, 1993).

The dentato-rubro-thalamic tracts are of particular interest in further elucidating the cerebellum's role in language processing. Dorsomedial dentate lesions produce the classical outflow tremor and ataxia, unlike disruption of pathways from the phylogenetically newer ventrolateral neodentate. These tracts travel by way of the medial ventrolateral thalamus to frontal neocortex areas 4 (motor), 6 (premotor/supplementary motor), and 8 (intellectual performance, initiative, and frontal eye fields directing contralateral fast saccades). Areas 4, 6, and 8 receive a majority of their dentate input from the dorsomedial portion of the nucleus. Fibers from the most medial portion of the ventrolateral nucleus feed "Broca's area," Brodman's 44 (encoding articulation), and the more prefrontal area 45 (word finding). Temporal lobe language areas project to the cerebellum by way of pontine nuclei. Frontal language areas project either by way of this pathway, by way of fronto-temporal fibers, or directly by way of fronto-pontine tracts (Figs. 3A and 3B).

The lateral cerebellum receives input from the frontal cerebrum in two ways. In one, mossy fibers project there from the pons. In the other, the inferior olivary nucleus (receiving cortical input by way of the red nucleus) sends climbing fibers to the cerebellum. This completes the eloquent loop system between the lateral cerebellum and the frontal cerebrum (Fig. 3B).

The exact cause of mutism in patients with these cerebellar injuries remains elusive. Exactly what the presence of pseudobulbar affect means anatomically and pathologically is unclear both in this syndrome and in our patient. Brainstem function and comprehension are intact, and this mutism occurs only after posterior fossa surgery, with prominent cerebellar speech during the recovery period. Neither meningitis, hydrocephalus (Ferrante, 1990; Salvati, 1991), vasospasm, nor stroke seem likely causative, since these do not occur uniformly with this syndrome. Likewise, neither postoperative dentate edema nor tracking brainstem edema (Volcan, 1986; Ammirati, 1989; Humphreys, 1989) occur universally with cerebellar mutism, and these entities would render impossible other functions these patients are able to perform.

Several authors point to lesions of the left more than right cerebellar hemisphere and vermis, involving the deep nuclei, as the anatomic site of the syndrome (Lechtenberg, 1978; Rekate, 1985; Humphreys, 1989). Bilateral stereotactic ablation or ischemia of the dentate nuclei has caused transient mutism (Fraioli, 1975; Guidetti, 1977; Ackermann, 1992), as has accidental injury of the nucleus interpositus (Siegfried, 1970). The likelihood of developing mutism my be proportionate to the extent of injury to the cerebellar midportion (vermis and paravermis) and dentate nuclei (Dietze, 1990; Dietze, 1991).

CONCLUSION

It appears that either vermian splitting or dentate destruction, discussed above, could account for the presented patient's mutism. That he was able to speak, when first seen by the medics, would suggest that the splitting of the inferior vermis is the more likely cause of the syndrome of cerebellar mutism, if we assume that the initial hemorrhage caused only dentate damage and that it was not until surgery that the vermis was incised. The postoperative MRI study shows dentate and vermian damage without brainstem injury. Because he remained intubated in the postoperative period, we cannot say whether or not there was a latent period of speech before the onset of his mutism (which we first discovered when he was later extubated). Perhaps mutism is the most severe form of dysarthria, the ultimate dyspraxia of voice coordination, rendering eloquence of speech impossible. Clearly the patient presented demonstrates that prolonged mutism can occur in adults with a non-neoplastic midline cerebellar lesion.

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