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The pathophysiology of oral pharyngeal apraxia and mutism following posterior fossa tumor resection in children

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✓ Mutism following posterior fossa tumor resection in pediatric patients has been previously recognized, although its pathophysiology remains unclear. A review of the available literature reveals 33 individuals with this condition, with only a few adults documented in the population. All of these patients had large midline posterior fossa tumors.

To better understand the incidence and anatomical substrate of this syndrome, the authors reviewed a 7-year series of 110 children who underwent a posterior fossa tumor resection. During that time, nine (8.2%) of the 110 children exhibited mutism postoperatively. They ranged from 2.5 to 20 years of age (mean 8.1 years) and became mute within 12 to 48 hours of surgery. The period of mutism lasted from 1.5 to 12 weeks after onset; all children had difficulty coordinating their oral pharyngeal musculature as manifested by postoperative drooling and inability to swallow. Further analysis of these cases revealed that all children had splitting of the entire inferior vermis at surgery, as confirmed on postoperative magnetic resonance studies. Lower cranial nerve function was intact in all nine patients.

Current concepts of cerebellar physiology emphasize the importance of the cerebellum in learning and language. The syndrome described resembles a loss of learned activities, or an apraxia, of the oral and pharyngeal musculature. To avoid the apraxia, therefore, the inferior vermis must be preserved. For large midline tumors that extend to the aqueduct, a combined approach through the fourth ventricle and a midvermis split may be used to avoid injuring the inferior vermis.

KEY WORDS • mutism • apraxia • vermis • cerebellum • posterior fossa tumor

SPEECH disturbances associated with cerebellar lesions have been previously recognized. From early descriptions of dysarthric patients who received cerebellar gunshot or shrapnel wounds²⁷ to the more recent research localizing speech dysfunction to the superior portion of the left cerebellar hemisphere,^{22,32} the importance of the cerebellum in the coordination of motor speech is clear. The syndrome of "posterior fossa mutism" has also received attention in recent literature. Over 30 case reports have appeared since 1980 linking postoperative mutism in the pediatric population with the removal of posterior fossa tumors.^{4,9,13,14,17,20,24,28,38,40,44,51} A few cases of the syndrome occurring in adults with midline posterior fossa lesions have also been described.^{13,44} Mechanisms implicated in the development of the syndrome have included bilateral destruction or edema of the cerebellar nuclei, vasospasm, postoperative meningitis, unrecognized hydrocephalus, and even psychiatric causes.

To better define the anatomical basis of postoperative mutism we reviewed all posterior fossa tumor resections that had been performed in our children's hospital over a 7-year period. During this time, nine children developed

an apraxia, or inability to execute complex motor movements of the oral pharyngeal musculature, as manifested postoperatively by mutism and difficulty in swallowing. In all these children, a complete splitting of the inferior vermis through the level of the pyramis had been performed and subsequently confirmed on postoperative magnetic resonance (MR) imaging. Surgical strategies to avoid the syndrome of oral pharyngeal apraxia and mutism are outlined based on this retrospective analysis.

Clinical Material and Methods

Pediatric Population

All children referred to the pediatric neurosurgical service at the Children's Hospital Medical Center, Seattle, Washington with the primary diagnosis of a posterior fossa tumor from January 1987 to December 1993 were included in the study. Those children referred for further therapeutic management after their initial posterior fossa craniectomy had been done elsewhere were excluded from this analysis. In addition, all children referred with

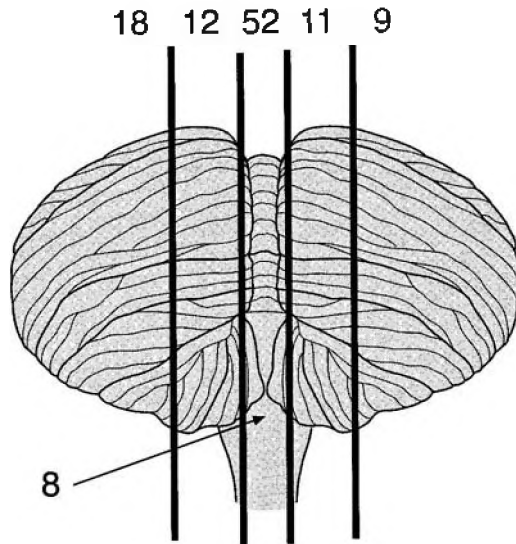


FIG. 1. Schematic representation of the posterior fossa showing the regions of the cerebellum divided sagittally and the corresponding number of patients who had their tumors approached through each region. The 52 patients with tumors of the vermis or fourth ventricle, and eight patients with tumors of the cervicomedullary junction form the group of midline lesions.

brainstem gliomas who did not undergo resection were excluded. Thus, only patients undergoing posterior fossa tumor resection at our institution were included in this retrospective study.

Data Evaluation

The study group was then evaluated based on data available from operative records and preoperative MR imaging or computerized tomography (CT) scans. The cerebellum was divided into six regions based on the epicenter of the tumor, presumed site of origin, and operative entry site (Fig. 1). Each cerebellar region was named according to which structures were split during the operative approach and grouped as follows: 1) vermis (all tumors located in the midline or within the fourth ventricle in which the vermis was split or retracted superiorly to facilitate tumor removal); 2) left and 3) right paramedian, respectively (all tumors in which the initial approach was through a cortical incision that was lateral to the vermis but within 2 cm of the fissure separating the vermis from the hemisphere); 4) left and 5) right hemisphere, respectively (all tumors approached through an incision lateral to those described for Groups 3 and 4); and 6) cervicomedullary (all dorsally exophytic brainstem lesions that displaced the vermis and tonsils superiorly).

The cases were then analyzed based on tumor pathology, patient age, region of surgical approach, and appearance of postoperative oral pharyngeal apraxia and mutism. The group of patients with midline tumors (Groups 1 and 6) were also examined to discover whether a split was performed in the vermis and to what level. To stratify this group, the vermis was divided into three zones, and the patients were categorized based on a determination of the level of vermis split. The superior vermis was defined as the lobules above the primary fissure (lingula, central lob-

TABLE 1
Location of lesions as classified by histological type in 110 children with tumors of the posterior fossa

Tumor Pathology	Location	
	Nonmidline	Midline
astrocytoma	28	20
medulloblastoma	12	28
ependymoma	3	9
glioma	5	2
other	2	1

ule, culmen), the middle vermis as those lobules from the primary fissure to the pyramidal fissure (declive, folium, and tuber), and the inferior vermis was located below the pyramidal fissure (pyramis, uvula, and nodulus).

Preoperative and postoperative MR images were examined in those patients who had midline tumors (Groups 1 and 6), because only children in these groups developed oral pharyngeal apraxia. The images were analyzed to determine the location of the split in the vermis and whether the split involved the entire inferior vermis. The number of children with oral pharyngeal apraxia and mutism was then determined in each group. Complete case records of the children with this postoperative syndrome were reviewed to determine age, sex, tumor pathology, duration and type of preoperative symptoms, length of mutism, association of postoperative infections or untreated hydrocephalus, and the time course of the return of speech function. Furthermore, CT scans obtained in the 48-hour postoperative period were reviewed for evidence of hemorrhage or infarction in any intracranial location.

Results

During the 7-year period from January 1987 through July 1993, 153 children with tumors located in the posterior fossa were referred to our service. Of the total number, 25 children had previously undergone surgery for the lesion, and they presented to us either with metastatic disease or with complications of their primary disease such as shunt malfunction. Another 18 patients had brainstem gliomas diagnosed by MR imaging and had subsequently received radiation therapy without a biopsy. The remaining 110 patients had a total of 117 posterior fossa craniectomies during this period.

The most common tumor type, juvenile pilocytic astrocytoma, comprised 48 (43.6%) of the cases; followed by medulloblastoma, 40 (36.3%); ependymoma, 12 (10.9%); other glioma, eight (7.3%); and miscellaneous cases, two (1.8%) that included an arteriovenous malformation and a rhabdoid tumor. The mean age of all children presenting for posterior fossa surgery was 6.2 years (range 6 months to 20 years).

The patients' brains were stratified into cerebellar regions based on tumor location and operative approach (Fig. 1). Regions 1 and 6 (52 and eight patients, respectively) were combined to create a group with midline tumors. The left and right paramedian groups had 12 and 11 cases, respectively, and the left and right cerebellar

Oral pharyngeal apraxia

TABLE 2

Relationship of oral pharyngeal apraxia and mutism with the level of vermis split in children undergoing surgery for tumors of the posterior fossa

Location of Vermis Split	Total	Oral Pharyngeal Apraxia With Mutism
none	17	0
inferior	23	5
middle	6	0
superior	7	0
inferior & middle	4	2
whole	3	2

hemisphere groups included 18 and nine children, respectively. As expected, medulloblastomas comprised the majority of midline tumors, whereas astrocytic tumors were found more frequently in nonmidline locations (Table 1).

Only children in the group with midline tumors developed oral pharyngeal apraxia and mutism postoperatively. Further analysis of the 60 midline tumors revealed that in 43 cases (72%) some split of the vermis was performed at the time of surgery, whereas 17 (28%) had no splitting of the vermis. The most common level of split involved the inferior vermis, as determined in 23 (54%) of the 43 patients included in this group. Splitting of the middle and superior vermis accounted for six (14%) and seven (16%) patients, respectively. Some overlap of the segmental splits occurred in four patients (9%) who had significant inferior and middle vermis splits, and three children (7%) in whom the entire vermis was split (Figs. 2 and 3). The postoperative syndrome of oral pharyngeal apraxia and mutism was only seen in children who had a vermian split that included the entire inferior portion of the vermis (Table 2).

Magnetic resonance confirmation of the level of the split was available in 39 (91%) of the 43 patients with splitting of the vermis, including 28 of the 30 patients who had inferior vermian splits. Review of the axial and sagittal T₁-weighted MR images allowed determination of whether the split involved the entire inferior vermis or just a portion of it (Table 3). In those children documented to have the entire inferior vermis split, nine (69%) of 13 children developed mutism. In the four individuals who had a complete split of the inferior vermis but did not have mutism, one was noted to be hypophonic with a reduced speech output during the 2 weeks following surgery, a second was found to be extremely irritable with aggressive outbursts of behavior, and the remaining two were noted to have minimal truncal ataxia. There were no cases of oral pharyngeal apraxia and mutism identified in the group of children undergoing tumor removal unless they also had a complete inferior vermian split extending through the anterosuperior portion of the inferior vermis including the deep white matter tracts of the corpus medullare.

Oral Pharyngeal Apraxia

Nine children with mutism and other signs of oral pharyngeal apraxia are described in detail in Table 4. These

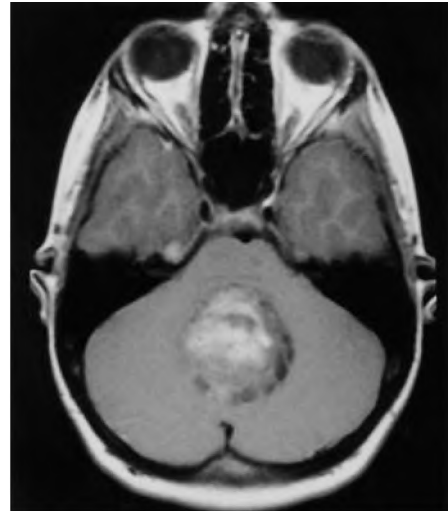


FIG. 2. Case 6. A 6-year-old girl with a split vermis who developed mutism postoperatively. A preoperative axial T₁-weighted magnetic resonance image with gadolinium enhancement shows a midline cystic lesion involving the vermis.

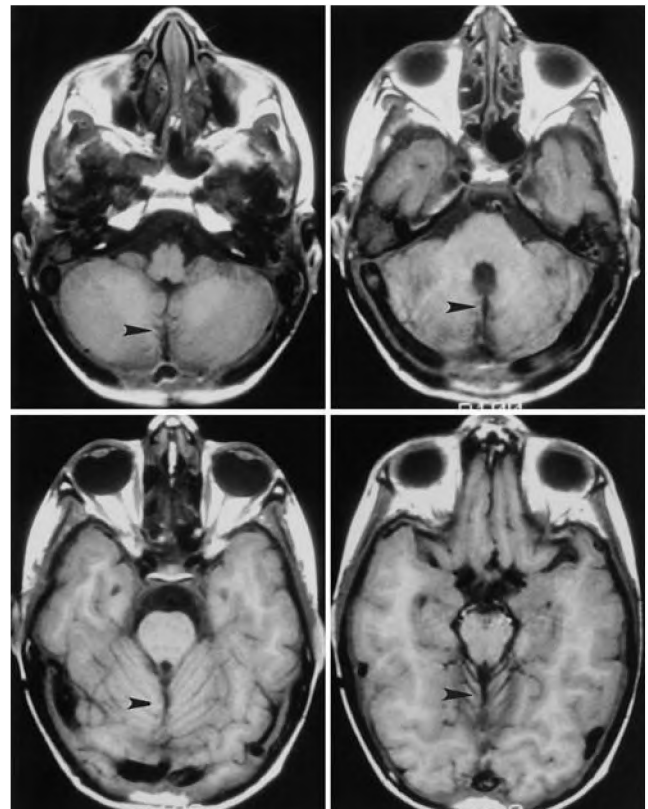


FIG. 3. Case 6. A series of postoperative axial T₁-weighted magnetic resonance images, caudal (upper left) to rostral (lower right) through the posterior fossa, showing the extent of the split in the vermis (arrows). There is no evidence of destruction of the paramedian nuclear structures.

TABLE 3

Relationship of the degree of inferior vermis split, as seen on postoperative magnetic resonance studies, and oral pharyngeal apraxia and mutism in 28 children undergoing surgery for tumors of the posterior fossa

Extent of Inferior Vermis Split	Total	Oral Pharyngeal Apraxia With Mutism
partial	15	0
entire	13	9

patients ranged from 2.5 to 20 years of age (mean 8.1 years). There were six girls and three boys. Six of the children had been diagnosed with medulloblastoma, again supporting the predominance of midline lesions as a component of the syndrome. Two children had astrocytomas and one had an ependymoma; eight presented with symptoms and signs of hydrocephalus and truncal ataxia, and one (Case 8) had undergone resection of a cerebellar astrocytoma 7 years prior to the current operation and presented again as a young adult demonstrating difficulty with upward gaze, gait ataxia, dysarthria, and a mild residual left hypotonia. None of the eight children presenting for the first time had evidence of cranial nerve palsies; one had evidence of pyramidal tract dysfunction (Case 2). Only one patient had preoperative evidence of a developmental speech delay (Case 6).

All patients underwent either MR imaging or CT scanning prior to surgery in addition to follow-up studies at 3- to 6-month intervals postoperatively to assess for tumor recurrence. Furthermore, all patients underwent CT scanning with and without contrast enhancement on postoperative Day 2 to evaluate the extent of resection and to rule out postoperative hemorrhage or infarct. All of these patients had partially or densely enhancing midline lesions blocking the fourth ventricle. One child (Case 1) had a lesion that appeared to arise from the roof of the fourth ventricle and extend posteriorly into the vermis, whereas the lesions of the remaining children appeared to expand and compress the vermis and plug the junction between the superior fourth ventricle and the aqueduct, thereby resulting in obstructive hydrocephalus. None of the patients had evidence of an infarct or hemorrhage to explain the postoperative deficits.

Every child, except one who was placed in the sitting

position at the time of surgery, underwent a posterior fossa craniectomy in the prone (Concorde) position after placement of a frontal ventriculostomy. The postoperative CT scans (performed within 48 hours after surgery) confirmed adequate ventricular catheter placement and drainage at the time of mutism onset. Gross total or near total (> 95%) resections using microscopic surgical techniques were performed in each case and verified with the postoperative imaging studies. At surgery, all children underwent splitting of the inferior vermis through the level of the pyramid. The division often extended to the caudal aspect of the middle vermis (folium and tuber) to fully visualize the aqueduct and allow for complete debulking. Four (44%) of nine tumors had "invaded," that is, were attached to, the floor of the fourth ventricle.

Except for one (Case 6) who appeared mute but was alert at the initial postoperative examination, all children awakened without deficits immediately following surgery. One required reintubation after a grand mal seizure 6 hours after surgery. All became mute within 48 hours of undergoing the surgery. In addition to the mutism, they had difficulty in swallowing and were unable to eat for a variable period of time after surgery. Testing of the lower cranial nerves showed an intact gag response and normal tongue movements on command in each child. Three of the children were able to resume oral intake within 1 week; however, the remaining six children needed to have tube feedings initiated. Swallowing evaluations suggested difficulty in preparing the food during the oral phase as well as inability to coordinate the pharyngeal phase, which resulted in delayed laryngeal elevation, and, ultimately, aspiration. Results of videofluoroscopic swallowing evaluations performed in two patients confirmed the difficulties in oral control and a subsequent delay in triggering the pharyngeal phase. The children regained the ability to eat, although in one girl (Case 3) this occurred nearly 1 year after the operation.

Mutism lasted from 1 to 12 weeks (mean 4.5 weeks) although residual dysarthria was detected up to 2 years after surgery in two of the children. Repeated evaluation of the dysarthria often showed diminished intelligibility as measured with the Computerized Assessment in Dysarthric Speakers (CAIDS) test, in addition to diminished stamina, measured as the mean length of utterance. Six children were admitted to the inpatient rehabilitation service for intensive speech and physical therapy. The children with medulloblastomas and ependymomas all

TABLE 4

Case summary of nine children with mutism and other signs of oral pharyngeal apraxia after undergoing surgery for tumors of the posterior fossa

Case No.	Age (yrs.) Sex	Pathology	Location of Vermis Split	Invasion of Brainstem	Time to Onset of Mutism	Length of Mutism	Length of Swallow Problems
1	4, M	ependymoma	inferior	yes	24 hrs	2 wks	5 days
2	12, F	medulloblastoma	inferior	no	immediate	1 wk	8 wks
3	9, F	medulloblastoma	whole	yes	12 hrs	4 wks	1 yr
4	6.75, F	medulloblastoma	whole	no	12 hrs	12 wks	8 wks
5	3, M	medulloblastoma	inferior	yes	24 hrs	1 1/2 wks	1 wk
6	6, F	astrocytoma	inferior-middle	no	24 hrs	6 wks	6 wks
7	2.5, M	medulloblastoma	inferior	yes	48 hrs	3 wks	5 days
8	20, F	recurrent astrocytoma	inferior-middle	no	12 hrs	8 wks	4 wks
9	10, F	medulloblastoma	inferior	no	60 hrs	3 wks	1 wk

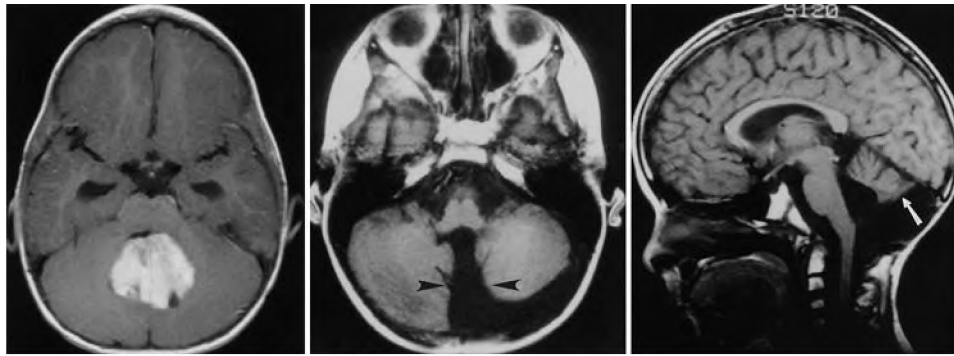


FIG. 4. Case 5. A 3-year-old boy with a split inferior vermis who developed postoperative mutism. *Left:* Preoperative T₁-weighted magnetic resonance (MR) image with gadolinium enhancement showing a diffusely enhancing midline lesion blocking the fourth ventricle. *Center:* Axial T₁-weighted MR image performed postoperatively revealing the split in the inferior vermis (*arrows*). *Right:* Sagittal T₁-weighted MR image showing the rostrocaudal extent of the split of midline structures. The split extends through the pyramis to the prepyramidal fissure(*arrow*).

received chemotherapy and radiation therapy protocols as outlined by the Children's Cancer Group study protocols.

Illustrative Case

Case 5

Examination. This 3-year-old boy presented with a 2-month history of morning nausea and vomiting. A physical examination revealed papilledema and gait ataxia. Radiological studies included a CT scan from the referring hospital and, subsequently, an MR study with and without gadolinium contrast material that demonstrated a large homogeneously enhancing lesion filling the fourth ventricle and compressing the vermis, resulting in obstructive hydrocephalus (Fig. 4 *left*).

Operation. Two days after admission the patient underwent a posterior fossa craniectomy following placement of a frontal ventriculostomy. During surgery, a soft, moderately vascular tumor was encountered upon laterally retracting the cerebellar tonsils. Histological examination confirmed that the lesion was a medulloblastoma. The inferior vermis was split for an approximate rostral to caudal distance of 1.5 cm. The tumor was attached to the right side of the floor of the fourth ventricle and a near-complete (99%) resection was achieved.

Postoperative Course. In the immediate postoperative period, the child was found to have a mild hypotonia of the left side; however, he was speaking in several-word phrases and could move all extremities when asked. On postoperative Day 3, he lost his ability to speak single words and appeared withdrawn from both his family and hospital staff. The postoperative CT scan showed no hemorrhage above or below the tentorium and the ventricles were decompressed by the ventriculostomy. Although he had an intact gag reflex, normal tongue protrusion, and full symmetrical facial movements, he was unable to eat or drink without aspirating. Therefore, a feeding tube was placed for 1 week postoperatively. By postoperative Day 10, he could again speak in single words, although these were difficult to understand. In addition, he was now able to swallow soft foods without aspirating. The child con-

tinued to improve and 2 weeks following surgery was transferred to the neurooncology service to begin chemotherapy and radiation treatments.

Throughout the course of treatment, he was closely followed by the rehabilitation and speech therapy services. Formal language testing 2 months postoperatively showed normal receptive language skills and vocabulary; however, the Goldman-Fristoc test of articulation demonstrated his speech to be only 75% accurate. The examiners noted a marked decrease in the vocal volume.

Follow-up MR studies obtained over the ensuing 2 years showed no recurrence of the tumor. Axial and sagittal T₁-weighted images revealed splitting of the inferior vermis through the level of the pyramis (Fig. 4 *center* and *right*). The last formal speech evaluation performed 18 months after surgery revealed expressive and receptive language beyond the child's age. At 1 year postoperatively, his articulation had returned to normal accuracy for his age.

Discussion

The role of the cerebellum in the coordinated production of speech has been recognized for some time. During World War I, Sir Gordon Holmes²⁷ observed over 40 patients with cerebellar wounds and noted that lesions of a single hemisphere often produced speech that was "slow, drawling and monotonous, but at the same time tends to be staccato and scanning." When the vermis was involved, he noted that disturbances of phonation and articulation were more pronounced; when both hemispheres were involved, speech became almost unintelligible. Holmes believed that extensive injuries to the cerebellum, particularly the vermis, led to an inability to coordinate all the separate movements that controlled the production of speech, a conclusion reiterated by other investigators.^{15,26}

More recently, Lechtenberg and Gilman³² provided a retrospective review of 162 patients with nondegenerative cerebellar diseases. They identified a subset of patients who had dysarthria and found that 22 of these 31 patients had significant left hemisphere destruction as their prima-

ry pathology. A subset of 26 patients had disease primarily in the vermis; however, only three of these patients developed a speech disorder. These authors concluded that the anterior paravermian region of the left cerebellar hemisphere was of primary importance in regulating the formation of speech.²² In addition, animal studies suggested that acoustic stimulation will result in the activation of the paravermian area of the left cerebellar hemisphere.³ Through a feed-forward loop connecting the cerebrum to the cerebellum,^{29,43} various parts of the cerebellum may monitor speech using the available auditory and sensory information and then adjust motor coordination accordingly. Thus, the importance of left cerebellar hemisphere damage in the dysarthric patient was determined.

Mutism is represented by a lack of voice or speech output in the awake patient, with the comprehension of speech and language remaining intact. It is not usually associated with cerebellar disease. Functional forms encountered in psychoses contribute to the majority of cases.⁵ However, several organic forms that can produce mutism are clearly documented and include the following: 1) lesions to portions of Broca's area, which produce what Benson⁵ describes as "aphemia;" 2) the superior mesencephalic artery syndrome resulting in damage to the reticular activating substance of the midbrain; 3) a dominant hemisphere supplementary motor area syndrome; 4) bilateral stereotactic thalamotomy in patients with Parkinson's disease; 5) diffuse bilateral subcortical infarcts; 6) the anterior opercular syndrome associated with buccomotor apraxia; and 7) lesions or infections of the vocal apparatus or nerves that innervate them.^{35,37,42,47}

Although the abnormalities of speech associated with cerebellar disease have been extensively categorized based on anatomical location, mutism was not included.¹² Some clinicians consider it the most severe form of dysarthria,^{24,38} or believe that it is indicative of the hypotonia often seen in cerebellar disease.²⁰ However, the presence of oral pharyngeal apraxia and mutism may indicate that the cerebellum is involved in higher functions such as learning, initiation, and cognitive processing, in addition to its role in adjusting motor output.^{35,36,46}

The first documentation of mutism following resection of a posterior fossa tumor appeared in 1979 when Hirsch, *et al.*,²⁵ were reviewing their outcome in a series of children with medulloblastomas. Thirty-three well-described cases (Table 5) have subsequently appeared in the English-language literature that link resection of cerebellar tumors and mutism (the "posterior fossa syndrome").^{4,9,13,14,17,20,24,28,38,40,44,51} Cochrane, *et al.*,¹⁰ in a report on all complications of posterior fossa tumor resections, found mutism occurring in six (5.7%) of 105 children following radical tumor removal. Although the syndrome has been primarily limited to children, a few adult cases have been published. However, two of these patients were only 20 years of age, while the third was 45 years of age and had a medulloblastoma that required a midline approach.^{13,44}

The common posterior fossa tumors of childhood have all been implicated, including medulloblastoma, astrocytoma, and ependymoma, in addition to a ruptured arteriovenous malformation. All children reported in the literature had midline lesions, and, in those cases where the surgical anatomy was carefully described (33% of report-

ed cases), all patients had their vermis split. The duration of complete speech loss ranged from 2 to 20 weeks with the majority of children having a 12- to 72-hour period immediately postoperatively in which speech was preserved. In nearly 30% of the cases, evidence of oral pharyngeal apraxia, as manifested by drooling, difficulty with swallowing, and aspiration, was present although no lower cranial nerve deficits were documented. Similarly, all nine of our cases had a period of time in which they were unable to swallow although none had lower cranial nerve deficits.

Wisoff and Epstein⁵² described seven children with similar problems after posterior fossa surgery, some of whom became mute; however, all of these children had well-documented cranial nerve deficits and the syndrome was termed "pseudobulbar palsy." These researchers believed the syndrome was caused by edema tracking through the middle and superior cerebellar peduncles into the brainstem. Because no lower cranial nerve deficits or cerebellar or brainstem edema were found on the postoperative imaging studies, our patients' symptoms resembled an apraxia of the oral pharyngeal apparatus more than a brainstem dysfunction.

Rekate and colleagues⁴⁰ described six children with extensive midline cerebellar tumors who developed mutism following resection. In their opinion, the dentate nuclei, which were the important structures damaged during the surgery, were responsible for the mutism. Bilateral injury to the dentate nuclei had first been reported as a cause of mutism by Fraioli and Guidetti¹⁹ who noted complete absence of speech in two patients who underwent bilateral stereotactic dentatolysis for dyskinesias. Other investigators have noted mutism with stereotactic lesions in the nucleus interpositus.⁴⁸ However, because only two of the 43 patients in Fraioli's series became mute, the role of the dentate nucleus as the sole anatomical substrate for oral pharyngeal apraxia remains uncertain.

Additional reports of "posterior fossa" mutism have also raised the possibility of postoperative edema resulting in bilateral dentate nuclei dysfunction.^{4,17,28,44} The majority of cases reported in the literature describe a period of intact speech immediately following surgery. Humphreys²⁸ claimed that a delayed response at 18 to 72 hours following surgery was the result of progressive swelling in the paramedian structures. Ammirati, *et al.*,⁴ documented midline swelling on postoperative CT scans that included the dentate nuclei. None of the children in our series had evidence of bilateral dentate nucleus edema or infarction on the postoperative CT scans or on follow-up MR imaging. Dietze and Mickle¹⁴ stated that injury to the ventrolateral portion of the dentate nucleus was responsible for interrupting the dentatocortical projections that link the planning of a motor event with its smooth production.^{21,34} Because these children still are able to perform other functions that diffuse dentate destruction or swelling would render impossible, another substrate besides the dentate nucleus is likely to be responsible for the syndrome.

Other factors have been implicated in mutism in addition to dentate destruction or edema.^{14,17,38,44} Some reports claim that mutism may be a functional disturbance reflecting a sense of betrayal in children whose parents subject them to difficult and painful operations. However, the

TABLE 5
Literature review of reports linking resection of cerebellar tumors and mutism*

Authors & Year	Case No.	Age (yrs), Sex	Pathology	Tumor Location	Vermis Split	Duration of Mutism (wks)	Long-Tract Sign	Swallowing Difficulty
Rekate, <i>et al.</i> , 1985	1	8, F	medulloblastoma	vermis	NM	6	rt hemiparesis	yes
	2	6, M	astrocytoma	vermis & lt hemisphere	NM	4	no	no
	3	3, NM	ependymoma	vermis	NM	8	NM	NM
	4	10, NM	medulloblastoma	vermis	NM	8	NM	NM
	5	9, NM	medulloblastoma	vermis	NM	12	NM	NM
	6	11, NM	medulloblastoma	vermis	NM	3	NM	NM
Volcan, <i>et al.</i> , 1986	1	8, F	medulloblastoma	vermis into 4th ventricle	yes	2	rt hemiparesis	no
Amirati, <i>et al.</i> , 1989	1	14, M	astrocytoma	vermis into 4th ventricle	yes	3	no	no
Humphreys, 1989	1	7, M	medulloblastoma	vermis & rt hemisphere	yes	20	no	yes
	2	3, M	medulloblastoma	4th ventricle	NM	7	yes	no
	3	7, M	medulloblastoma	vermis	NM	10	no	yes
	4	4.5, M	astrocytoma	cerebellar peduncle	NM	7	rt hemiparesis	NM
	5	10, F	ependymoma	vermis into 4th ventricle	yes	10	no	yes
Yonemasu (in Humphreys), 1989	1&2	NM	ependymoma	vermis	NM	4-12	NM	NM
	3&4	NM	medulloblastoma	vermis	NM	4-12	NM	NM
Dietze & Mickle, 1990-91	1	7, M	medulloblastoma	vermis & both hemispheres	yes	<6	no	no
	2	15, F	AVM hemorrhage	midline	NM	<12	no	no
Ferrante, <i>et al.</i> , 1990	1	9, F	astrocytoma	vermis	yes	4	no	yes
	2	5.5, F	astrocytoma	vermis	yes	8	no	yes
	3	6, F	astrocytoma	vermis	yes	8	no	yes
Gaskill & Marlin, 1991	1	8, F	medulloblastoma	vermis into 4th ventricle	NM	2	no	no
	2	7, M	medulloblastoma	vermis	yes	NM	NM	NM
	3	10, F	ependymoma	4th ventricle	NM	NM	NM	NM
Herb & Thyen, 1991	1	9, M	medulloblastoma	vermis	NM	10	rt hemiparesis	no
Nagatani, <i>et al.</i> , 1991	1	4, F	medulloblastoma	vermis	yes	10	no	yes
Salvati, <i>et al.</i> , 1991	1	20, M	medulloblastoma	vermis into 4th ventricle	NM	4	no	no
Catsman-Berrevoets, <i>et al.</i> , 1992	1	6, M	medulloblastoma	vermis	NM	7	rt hemiparesis	yes
	2	8, F	medulloblastoma	vermis	NM	8	lt hemiparesis	NM
	3	8, M	medulloblastoma	vermis	NM	8.5	no	yes
D'Avano, <i>et al.</i> , 1993	1	20, F	medulloblastoma	midline	NM	6	no	NM
	2	45, M	medulloblastoma	midline & rt hemisphere	NM	8	no	NM

* NM = not mentioned in the literature; AVM = arteriovenous malformation.

physical basis for this may be postoperative edema or ischemia produced by spasm of the cerebellar vessels. Finally, some authors have stressed the importance of postoperative hydrocephalus or meningitis.^{17,44} In our series, none of the children had bacterial meningitis and all had adequate ventricular drainage as confirmed by postoperative CT scans.

Although these mechanisms have been offered to explain the "posterior fossa mutism" syndrome, the anatomical basis for the syndrome remains difficult to define. The pyramis and uvula appear to be the areas crucial to the development of this syndrome and are included in the spinocerebellum, which is believed to be primarily responsible for the maintenance of posture and truncal stance.^{7,22} However, sensory stimulation provides a somatotopic map of the cerebellum in which this region receives tactile information not only from the limbs, but also the head and face.^{7,21} Nontactile stimulation from auditory and visual sources is also represented in the superior portion of the inferior vermis.⁴⁹ These areas also receive information from the motor and sensory cortices of the cerebrum as well as subcortical areas that are responsible for speech initiation.² Thus, surgical damage to these posterior paravermian and vermian regions may uncouple cerebellar control from cortical output, thus

interfering with smooth motor execution of oral pharyngeal functions. In addition, primate studies have confirmed that midline cerebellar structures participate in the coordination of laryngeal and respiratory functions necessary for the smooth production of speech.³¹

Midline cerebellar cortical structures have complex nuclear-cortical and cortical-nuclear projections that link the paravermis and the vermis with the cerebellar nuclei.⁵⁰ These nuclei, in turn, have important connections with the pontine nuclei, the ventral lateral thalamus, and subsequently, motor and sensory association areas such as the supplementary motor area that have proven necessary for the initiation of speech.^{7,8} The paravermian region uses sensory information from the spinocerebellar and cuneocerebellar tracts to compare learned motor programs with intended cerebral output, and adjusts final motor output accordingly.^{2,30,36} If the cerebellar regions receiving sensory input from the oral pharynx were damaged, either directly or through postoperative edema, the motor output (speech and swallowing) could be adversely affected.

At the time of surgery all of the children at our institution with oral pharyngeal apraxia had splitting of the deep inferior cerebellar white matter (corpus medullare cerebelli). Although no cerebellar transcommissural tracts were previously believed to be of physiological signifi-

cance,^{7,8,21} researchers have suggested that coordinating complex movements requiring bilateral musculature must involve both cerebellar hemispheres acting in unison. Pathways do exist that connect adjacent lobules as well as the inferior lateral sections of each hemisphere.¹⁶ These areas of neocerebellum are the phylogenetically newest regions of the cerebellum and therefore should be responsible for higher human functions such as speech and ideation.^{1,33,36} Anatomical studies in cats show a transcommissural system with pontine nuclei exhibiting crossed collaterals that project to the neocerebellum.⁴¹ Afferents to these pontine nuclei project from a variety of cerebral cortical areas with important contributions from the pre-motor association and supplementary motor areas. Just as damage to these cortical regions may result in prolonged periods of buccomotor apraxia, interrupting the cerebellar portion of the loop by transecting the transcommissural pathway may lead to the syndrome of oral pharyngeal apraxia following posterior fossa tumor resection.^{23,37}

Evidence obtained using positron emission tomography and single-photon emission computerized tomography supports the importance of bilateral cerebellar activation during speech. These techniques have revealed bilateral posterior paravermian activation during speech as well as bilateral activation of the inferolateral cerebellar hemispheres, with word generation and motor imagery tasks that are the prelude to speech.^{39,43} Another clinical correlate supporting the role of the midline cerebellum in speech has been observed in the MR images from children with autism or the fragile X syndrome. These children, who have had developmental and speech delays, show a volume loss in the cerebellum, particularly the posterior vermis.¹¹ The association of delayed speech and underdevelopment of the posterior vermis emphasizes the importance of this region in the acquisition of speech skills.

Apraxia may be defined as a disorder in the understanding of complex motor commands and the execution of certain learned movements.⁶ Although the parietal lobe has traditionally been associated with these deficits, evidence indicates that the cerebellum contributes to learning and the execution of motor programs. Experimental evidence supports the role of the cerebellum in learned responses such as the nictitating membrane response and the acoustic startle response,^{30,34} while clinical studies in humans with cerebellar hemisphere atrophy confirm that these people have difficulty in learning motor tasks.⁴⁵

A recent report by Fiez, *et al.*,¹⁸ expanded this view by describing an individual who had impaired nonmotor learning while performing cognitive tasks following a cerebellar stroke. The authors conclude that the cerebellum may be an important structure in a long-term memory or learning system that is distinct from the mesial temporal structures. As a result, it appears that the cerebellum is important not only for motor execution but also for planning and initiation of learned mechanisms. After splitting the cerebellar hemispheres, children with oral pharyngeal apraxia may initially not be able to coordinate complex learned movements that require simultaneous bilateral activation. The fact that speech returns attests to the ability of the cerebellum to participate in complex, coordinated oral pharyngeal motor tasks critical for phonation and swallowing.

Conclusions

Radical splitting of the inferior vermis may result in a syndrome of oral pharyngeal motor apraxia with postoperative difficulty in speech and swallowing. The precise mechanisms through which the cerebellum is involved in the development and maintenance of speech remain difficult to clarify, although this study implies a critical role for the inferior vermis in this postoperative syndrome. However, recognition of the syndrome will help warn families preoperatively of this potential set of distressing yet reversible postoperative symptoms. We limit the split in the inferior vermis to less than 1 cm, and use a split above the prepyramidal fissure (the middle vermis), should higher access to the fourth ventricle be required. Postoperative MR imaging is useful to rule out noncerebellar causes of mutism and to confirm the extent of the inferior vermis split.

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Oral pharyngeal apraxia

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