

Cerebellar Mutism after Posterior Fossa Surgery

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Abstract. A large midline posterior fossa medulluloblastoma and a cerebellar arteriovenous malformation with associated clot were moved with postoperative mutism. Mutism was immediate and resolved into an ataxic dysarthria with residual mild dysarthria at 3 months. The literature is reviewed, and the clinicopathological and neurophysiological data of cerebellar function in speech are discussed. Acute injury to the midportion of the cerebellum with or without dentate nuclear involvement can cause a spectrum of speech disturbances. The more widespread the injury to the midportion of the cerebellum with dentate nuclei involvement the greater the risk of developing postoperative mutism.

Introduction

The cerebellum has been associated with a wide range of speech dysarthrias. These dysarthrias range from a monotonous, scanning speech to an explosive, hesitant outburst of speech [1, 2]. Degenerative disorders of the cerebellum have most commonly been associated with speech abnormalities, but diffuse or widespread injury by hemorrhage, infection, or tumor have also been associated with speech abnormalities.

Mutism can be defined as the inability to produce verbal output in a cognitively alert patient with the ability to read and write with intact comprehension and no evidence of oral apraxia. Posterior fossa lesions causing speech abnormalities have been attributed to brain stem, cranial nerve, and cerebellar pathologies. Brain stem and cranial nerve pathologies causing mutism are usually associated with other neurologic signs, i.e., long-tract deficits or decreased level of consciousness, and respiratory compromise, respectively.

Transient mutism was recently observed in 2 of our patients after posterior fossa surgery. Our 2nd case involving an arteriovenous malformation is unique because all cases reported have involved tumors in the

posterior fossa. This rare phenomenon after posterior fossa surgery is a dramatic deficit and should be included in the preoperative discussion with the patient and family. We propose a hypothesis for mutism as a result of cerebellar pathology based on our experience with these 2 cases.

Case Reports

Case 1

A 7-year-old presented with a chief complaint of severe frontal headaches associated with intermittent right-arm pain for a duration of approximately 2 months. His physical examination was pertinent for mild truncal ataxia and normal speech production.

Preoperative magnetic resonance imaging (MRI) of the head revealed a large midline posterior fossa tumor with moderate obstructive hydrocephalus (fig. 1, 2). The patient underwent a suboccipital craniectomy and C1-2 laminectomies in a left-sided modified park bench position. Intraoperative findings revealed chronic tonsillar herniation and a widened, thickened vermis. Just beneath the vermicular cortex, a soft, greyish tumor was encountered. The tumor was followed laterally into both cerebellar hemispheres and anteriorly until the floor of the fourth ventricle was encountered. The tumor was removed except for a small attachment to the floor of the fourth ventricle. The pathological diagnosis was medulloblastoma.

The patient tolerated the procedure well and was placed in the

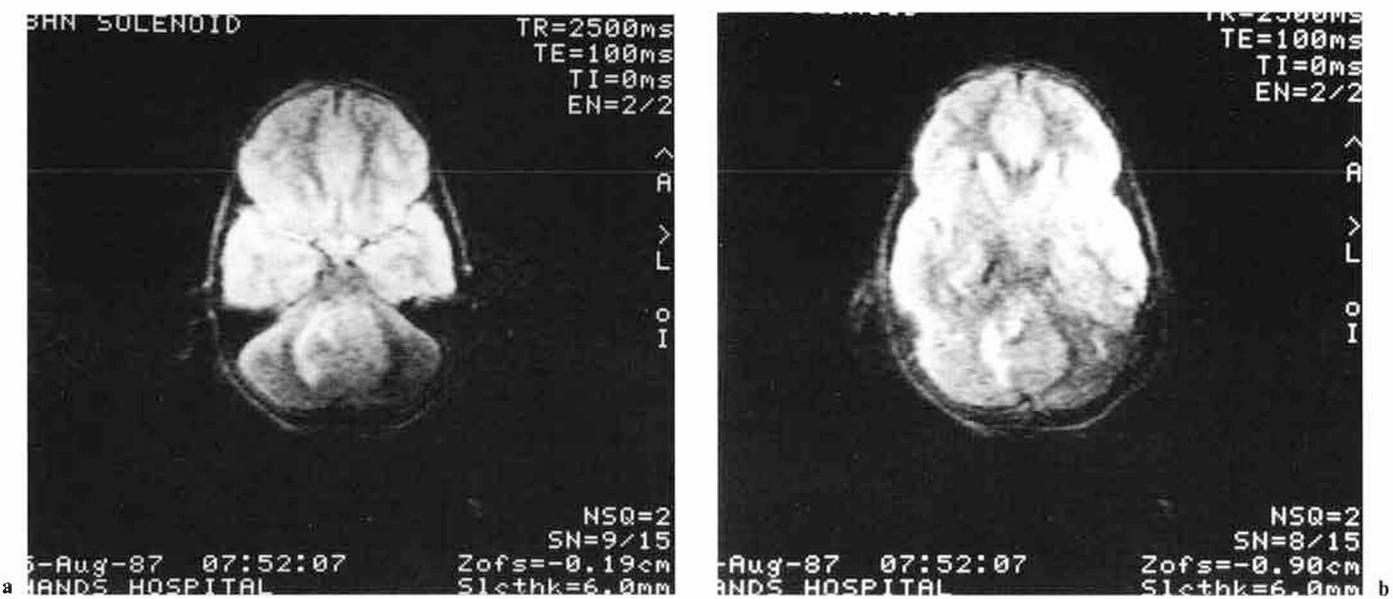


Fig. 1. Transaxial MRI sections with T2-weighted images demonstrating a large midline posterior fossa tumor. **a** Entire midportion of the vermis involved with obliteration of the fourth ventricle. **b** Tumor encroaching on the tentorial incisura with left superior paravermal involvement.

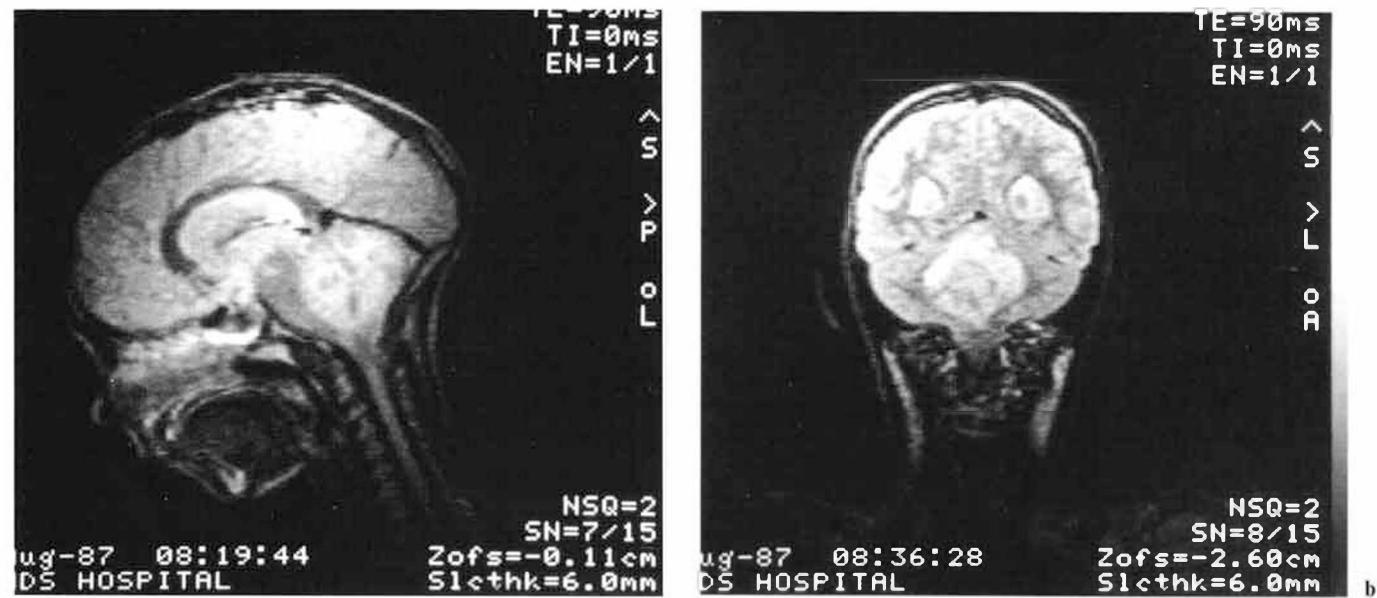


Fig. 2. Midsagittal (a) and coronal (b) MRI sections with T1-weighted images demonstrating obliteration of the fourth ventricle by a large tumor extending superiorly, involving lingula, central lobule, and culmen, and posteriorly, involving declive, folium, and tuber of the vermis.

pediatric intensive care unit. He was bright, awake and alert, and neurologically intact with moderate truncal ataxia and mild right-arm ataxia. He was mute with rare incomprehensible sounds, but was able to read and write with good comprehension and had no evidence of oral apraxia.

He was treated with spinal and cranial external-beam radiation.

Routine clinic follow-up at 6 weeks revealed resolution of his appendicular ataxia, but with persistence of truncal ataxia and moderate dysarthria. His dysarthria was characterized by a sparse, slow, and monotonous speech associated with intermittent explosive-hesitant outbursts and exaggerated facial grimacing. At 3 months of follow-up the patient had almost complete resolution of his dysarthria. MRI

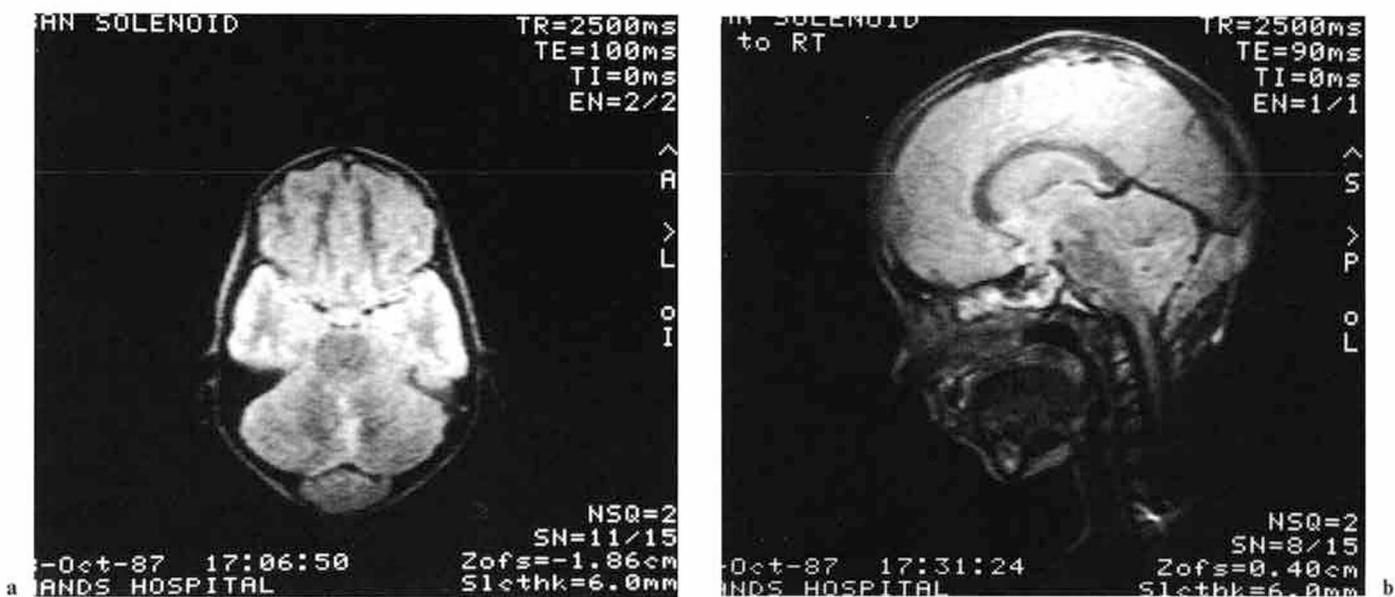


Fig. 3. Postoperative MRI sections demonstrating encephalomalacia of inferior and middle vermis and paravermis. **a** Transaxial T1-weighted image. **b** midsagittal T1-weighted image.

6 months postoperatively showed encephalomalacia in the posterior fossa around the fourth ventricle and no tumor (fig. 3).

Case 2

A 15-year-old awakened at 0.400 h with an acute severe headache. Her headache was associated with photophobia and bilateral ear pain. She deteriorated rapidly with loss of consciousness and respiratory compromise. At a nearby hospital an emergent computerized tomography (CT) scan showed acute posterior fossa bleed and acute obstructive hydrocephalus. An emergent suboccipital craniectomy and hematoma evacuation with placement of a ventriculostomy was performed. She awakened slowly over several days. Examination revealed diffuse hypotonia, bilateral appendicular ataxia with marked truncal ataxia, and mutism. Placement of a right frontal ventriculoperitoneal shunt was performed, and she was transferred to the Shands Hospital at the University of Florida.

A CT scan showed inhomogeneous areas of lucency within both cerebellar hemispheres adjacent and posterior to the fourth ventricle (fig. 4). No substantial mass effect was noted. Cerebral angiography revealed a superior vermian arteriovenous malformation principally fed by the left superior cerebellar and the right posterior inferior cerebellar arteries and minimally by the right superior cerebellar artery (fig. 5). The patient underwent exploration of the suboccipital defect in a left modified park bench position. The right posterior inferior cerebellar artery and the left superior cerebellar artery were identified and followed to the superior vermal and paravermal nidus. The nidus was posterior to the fourth ventricle and extended into the area of the deep cerebellar nuclei. A complete resection of the arteriovenous malformation was performed without complications. Her postoperative course was unremarkable with no new neurologic deficits. The postoperative CT scan showed cerebellar encephalomalacia of the bilateral paravermal hemispheres and the middle superior vermis (fig. 6). The ventricular size was normal.



Fig. 4. Transaxial CT section with intravenous contrast enhancement showing irregular enhancement of the midvermis and blotchy areas of lucency bilateral in the paravermal zones.

The patient was sent for inpatient rehabilitation. At 3 months she was able to produce all sounds, but had a monotonous, slightly labored, and bradykinetic dysarthria. Her appendicular ataxia had almost completely resolved, but she persisted to have moderate truncal ataxia requiring mild assistance for ambulation.

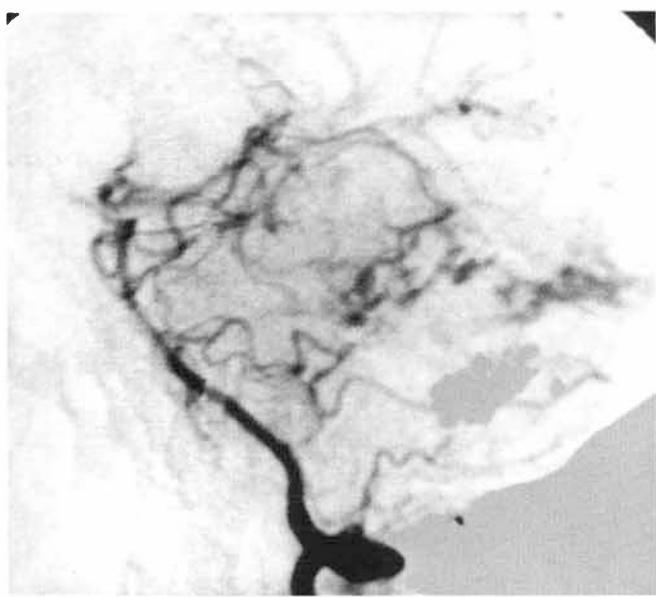


Fig. 5. Lateral vertebral angiography demonstrating a posterosuperior vermian arteriovenous malformation nidus principally fed by the left superior cerebellar and the right posterior inferior cerebellar arteries, extending intraparenchymally adjacent to the dentate nuclei.



Fig. 6. Postoperative transaxial CT section demonstrating encephalomalacia of the posterosuperior vermis and the adjacent paravermal zones.

Discussion

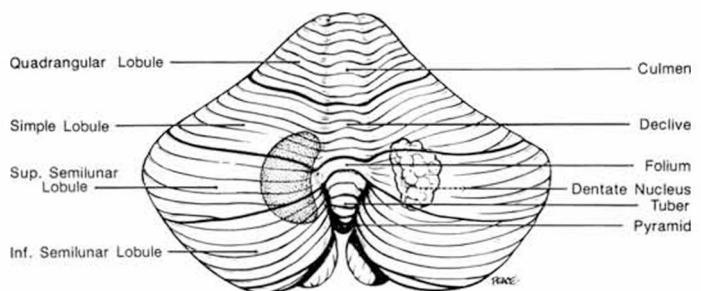
A review of the literature disclosed 19 cases of transient mutism after surgical removal of posterior fossa tumors. Rekate et al. [3] reported 6 cases of children with mutism which lasted 1–3 months. All of the children were severely dysarthric during recovery. Volcan et al. [4] reported a case of transient mutism after surgical removal of a posterior fossa tumor. Humphreys [5] reported 5 cases of postoperative cerebellar mutism in children recovering from posterior fossa surgery. This author comments that 3 of his cases had delayed onset of mutism. These delayed cases were associated with postoperative neurologic decline, except 1 case who had documented residual hydrocephalus requiring shunting 2 weeks later. These cases still had resolution into a cerebellar dysarthria with complete or near complete recovery. Humphreys [5] also comments on 4 cases reported by Yonemasu with 'giant' fourth ventricular tumors. These patients developed postoperative mutism which occurred after a brief interval of normal speech performance. The mutism resolved into a cerebellar dysarthria within 1–3 months and continued to complete or near complete resolution. Ferrante et al. [6] most recently reported 3 cases of transient mutism after surgical removal of large midline cerebellar pilocytic astrocytomas. The mutism in their cases evolved after a 'clear' interval of speech production of 36–48 h. The cases reported of transient mutism with a clear interval involve a secondary insult such as meningitis or hydrocephalus. A synopsis of the pertinent clinical characteristics of these cases and our 2 cases are listed in table 1.

Postoperative evaluation of this complication included speech therapy, clinical psychology, CT, elec-

Table 1. Synopsis of the reported cases (including our 2)

<i>Age, years</i>	
Range	2–15
Mean	7.5
<i>Diagnoses</i>	
Large midline 4th ventricle tumors	
Medulloblastomas	11
Ependymomas	4
Cystic astrocytomas	5
Total	20
Vascular malformation with hemorrhage	1
<i>Duration of mutism</i>	
Range from	3 weeks to 7 months
Mean	3 months

Fig. 7. Schematic illustration showing the posterosuperior surfaces of the cerebellum. The stippled area in the left paravermal region of simple lobule and superior and inferior semilunar lobules is the proposed area involved in speech disturbances from cerebellar pathology and with extension into the dentate nucleus possibly in cerebellar mutism.



troencephalography, indirect laryngoscopy, brain stem evoked response potentials, carotid ultrasound, and spinal myelography. Speech evaluation and indirect laryngoscopy showed no evidence of oropharyngeal or laryngeal musculature abnormalities or vocal cord abnormalities. Clinical psychology could not support a diagnosis of depression and conversion reaction. Evoked potential responses revealed no brain stem abnormalities, and electroencephalography revealed no seizure activity. Spinal myelogram and carotid Doppler studies were normal, and a CT scan revealed expected postoperative changes. Thus, the conclusion of the extensive evaluations was that the mutism was of cerebellar origin.

The association between cerebellar lesions and defects of speech production has long been recognized and reported with a multitude of cerebellar diseases. In 1823 Flourens described the tremor, staggering gait, and slurred speech accompanying cerebellar disease. In 1917 Holmes [7] reported that speech associated with gunshot wounds to the cerebellum was slow, drawling, and monotonous with an unnatural separation of the syllables and a jerky, often explosive articulation. This author concluded that speech production was more seriously affected when vermis and mesial hemispheres of the lateral lobes of the cerebellum were injured. Speech in bilateral cerebellar lesions in 2 patients was so indistinct that it was scarcely intelligible [7]. Homes [8] in his classic article for the Croonian Lectures in 1922 reiterated these observations, but also commented that disease limited to one lateral lobe could affect speech production.

Amici et al. [9] reviewed 250 patients with cerebellar tumors prior to surgery and found the highest incidence of dysarthria with lesions in paravermal and lateral elements of the hemispheres. Multiple studies reporting on cerebellar hemorrhages implicate unilateral left-sided lesions associated with speech disturbances [10-14]. Gilman and Lechtenberg [12] and Lechtenberg and Gilman

[15] present good evidence in nondegenerative diseases of the cerebellum that damage to the superior portion of the left paravermal cerebellar hemisphere is associated with disturbance of speech production (fig. 7). Neurophysiologic studies with auditory and laryngeal nerve stimulation show inputs into this superior paravermal segment of the left hemisphere [16-18].

The clinicopathological and neurophysiological data localize speech modulation to the midportion of the left paravermal area. Kent and Netsell [1] have described the role of the cerebellum in speech as to supplement and revise as necessary the basic cortically generated motor program by monitoring the positions of the speech organs and generating appropriate motor instructions to realize the articulatory targets. With respect to the longitudinal-zonal organization of the cerebellum, the paravermal zone is involved in a closed sensorimotor-cerebellar-sensorimotor loop capable of updating the cortically generated programmed movement based on the integration of central and peripheral inputs [19].

In this extensive investigation of disturbances in cerebellar lesions mutism is not described. However, mutism of cerebellar origin cannot be discarded. Fraioli and Guidetti [20] in their report on the effects of stereotactic lesions of the dentate nucleus for dyskinetic syndromes report 2 cases of transient mutism as a complication (for 1 and 3 months, respectively). Tolbert et al. [21] presented evidence that the dentate nucleus has nucleocortical projections over large areas of the ipsilateral cerebellar cortex, including the vermis, and that the dentato-nucleocortical projection to the mesial cerebellar hemispheres was large as compared with the nucleocortical projections from the interposed and fastigial nuclei. The implication of this evidence is that the higher primate cerebellum functionally may not conform to the longitudinal-zonal organization and that the dentate efferent output would be expected to affect cerebellar cortical integration not only in the neocerebellum but also in the paleocerebellum. Morphological-

ly, the dentate can be divided into a ventrolateral region, the neodentate, and a posteromedial region, the paleodentate.

Brown [22], with reference to the paper by Snider and Eldred [23], divides the cerebellum into major functional divisions based on cerebrocerebellar relationships. The midportion of the cerebellum receives input from auditory and visual sources. This region correlates with the clinicopathological and neurophysiological data presented for speech modulation.

If speech is considered a fine-skilled movement requiring the coordination of the bilateral axial musculature, then one may hypothesize that the dentate nucleus can integrate this motor production. The paleodentate having interconnections with vermis and paravermis could modulate these axial musculatures for the anticipated preprogrammed motor command for speech generated by the cerebral cortex. Mutism, however, still must be explained. If the cerebellum does provide a memory of learned movement to increase efficiency and execution, then, perhaps, an acute shut down of this system could prevent performance of the learned movements by the motor system [24, 25].

Conclusion

Mutism as an apparent result of cerebellar injury has been documented in 19 cases plus our 2 cases. This mutism is not restricted to tumor pathology, but may be associated with any acute pathological insult to the midportion of the cerebellum. Mutism is a failure to produce comprehensible sounds in a cognitively alert patient with apparent normal functional ability of lips, tongue, and laryngeal musculature, though there may be an associated dysphagia. The mutism resolves completely or to a mild cerebellar dysarthria over a 3-month period. The failure to produce speech appears associated with the inability to coordinate articulatory postures in order to produce enough subglottic pressure for the production of sound.

The midportion of the cerebellum is at risk of injury in midline fourth ventricular tumors, especially midline medulloblastomas. Review of the microsurgical anatomy shows the close relationships of the superior medullary velum, the cerebellar peduncles, and the midportion of the vermis (declive, folium, and tuber) [26]. The adjacent paravermis consists of the simple lobule and the superior and inferior semilunar lobules. The midportion of the cerebellum is generally involved in fourth

ventricular tumors, and if the vector force of tumor growth extends to the inferior part of the roof of the fourth ventricle, the dentate nucleus may be involved. Dandy [27] warns to carefully avoid the dentate nuclei upon splitting the vermis. The dentate nuclei are located just rostral to the superior pole of the tonsils and are wrapped around the superolateral recess of the ventricle near the inferior medullary velum.

Acute injury to the midportion of the cerebellum with or without dentate nuclei involvement can be responsible for a spectrum of speech disturbances. The more widespread the injury to the midportion of the cerebellum with dentate nuclei involvement, the greater the risk of developing postoperative mutism. Mutism is such an overwhelming deficit that this possible complication after posterior fossa surgery should be discussed as part of the preoperative planning.

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Editorial Comment

With the recent reports of the uncommon problem of mutism appearing after posterior fossa surgery, neurosurgeons feel somewhat relieved that the disorder is recognized (even though its explanation is still lacking) and, most of all, that recovery can be anticipated.

The 2 cases reported here are in the first instance typical of those documented earlier in that a large midline posterior fossa tumor was responsible for the patient's postoperative complication. The second case is more intriguing to me as this is the first to my knowledge in which a ruptured cerebellar arteriovenous malformation

has allegedly caused the mutism which in turn was detected prior to any operative intervention. In neither instance was there another parallel complication (infection, hydrocephalus, postoperative hematoma) which must in all of these cases be sought and remedied.

This second case provides a convincing argument for disruption of paravermian tissues as being the anatomic substrate for the 'cerebellar mutism'. The authors have explored that matter in detail. Yet consider all of the pathological processes and surgical incursions in the cerebellar midline, and just how infrequent mutism actually is. Why?

Robin P. Humphreys, MD