Transient Cerebellar Mutism after Posterior Fossa Surgery in Children

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CEREBELLAR MUTISM HAS been reported after surgery for posterior fossa tumors in children and, rarely, in adults. The pathogenesis of this syndrome remains unclear, and controversy exists regarding whether it is a purely psychogenic disorder or an organic syndrome. The anatomical substrate for the mutism also remains unknown. We encountered five cases of postoperative transient cerebellar mutism in a consecutive series of 63 children with posterior fossa tumors. These cases were analyzed and compared with the patients without mutism to find predictive factors for the occurrence of mutism, with the hope of elucidating further the pathophysiological mechanism. The most significant finding was the presence in all cases of a period of cerebellar dysarthria after resolution of the muteness. We, therefore, believe that cerebellar mutism is an extreme form of dysarthria, rather than a real cognitive deficit or a psychological disturbance. (Neurosurgery 37:894–898, 1995)

Key words: Brain neoplasm, Cerebellum, Child, Hydrocephalus, Mutism, Posterior fossa

Mutism, defined as the total absence of speech and sound in an awake and conscious patient, was recognized as a complication of surgery for posterior fossa tumors in children by Rekate et al. in 1985 (26). Previous authors had noticed similar syndromes, e.g., the "pseudobulbar palsy," described by Wisoff and Epstein (31) or had mentioned mutism as a symptom in a larger syndrome of "retardation" (17). Muteness occurring after stereotactic lesions in the dentate nucleus has been known since 1975 (14).

Since the description by Rekate, some 48 patients with this form of cerebellar mutism have been reported. These reports have generally been anecdotal, and although much controversy has arisen concerning the pathogenesis of the loss of speech, no predictive factors for the syndrome have been described. Because these factors might shed some light on the pathophysiology, we analyzed our series of children with posterior fossa tumors and compared these data with the other published cases.

PATIENTS AND METHODS

A retrospective analysis was made of a consecutive series of 63 patients, aged less than 16 years, who underwent surgery for a tumor in the posterior fossa during a 10-year period (January 1984 to December 1993). In five of these patients, postoperative muteness occurred. The case records, operative reports, and pre- and postoperative computed tomographic (CT) and magnetic resonance imaging scans were scrutinized for possible factors that could influence the postoperative loss of speech. All observations described in the literature were analyzed, including those cases only described in abstract form, to find common denominators for the syndrome. Statistical analysis was performed with χ^2 and Student's t tests.

RESULTS

Postoperative mutism occurred in 5 of the 63 patients. Some general clinical characteristics of these patients are described in *Table 1*. There were three girls and two boys, with ages ranging from 41 to 152 months. A period with normal speech before the mutism was found in only one patient; in the second patient, the mutism was noted after a period of coma and stupor of less than 48 hours; and in the three other children, the mutism was present at the time of the first neurological examination after surgery. The duration of complete absence of speech ranged from 14 to 76 days, and it was followed in all five children by a period with moderate to severe dysarthria. A comparison between the characteristics of the 5 mute patients and the 58 other patients is summarized in *Table 2*.

TABLE 1. Clinical Characteristics in the Five Patients with Cerebellar Mutism

Patient No.	Sex	Age (mo)	Nonmute Period	Days to Recovery	Dysarthria
1	F	152	0	16	Yes
2	F	70	0	76	Yes
3	M	41	0	63	Yes
4	F	64	7 d	14	Yes
5	M	83	0	28	Yes

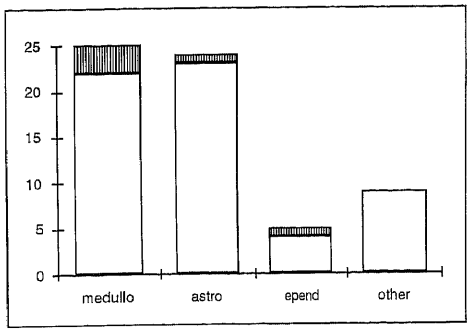

FIGURE 1. Graph illustrates the histological findings of the cases with (*shaded*) and without (*white*) mutism. *Medullo*, medulloblastoma; *astro*, astrocytoma; *epend*, ependymoma.

The mean age did not differ significantly, nor did the sex distribution. The clinical symptoms at presentation, the occurrence of hydrocephalus, and the largest diameter of the tumor on the preoperative CT scan did not differ significantly. The presence of papilledema was significantly associated with postoperative mutism ($P < 0.05$). Three of the 23 lesions located in the vermis and 2 of the 19 located in the vermis with an extension toward the hemispheres showed postoperative mutism. None of the 13 patients with a purely hemispheric tumor had mutism, nor did any of the five patients with tumors predominantly affecting the brain stem.

The histological findings are presented in *Figure 1*. There were 25 cases of medulloblastoma (3 cases of mutism), 24 astrocytic tumors (1 case of mutism), 5 ependymomas (1 case of mutism), and 9 other tumor types (dermoid, epidermoid, cavernoma, hemangioblastoma).

Brain stem invasion or involvement was observed in 4 of the 5 patients with mutism and in 18 of the other 58 patients. This reached significance at the 0.05 level. In these four patients, the surgical resection was subtotal, leaving a small layer of tumor in the floor of the fourth ventricle. Mutism occurred only once in the patients in whom the tumor was resected radically.

Cranial nerve deficits were not significantly more frequent in the patients suffering from cerebellar mutism: oculomotor disturbances were found in 25 of 58 patients versus 4 of 5 in the mute group; facial nerve paresis occurred in 9 of 58 patients versus 3 of 5; and lower cranial nerve dysfunction occurred in 4 of 58 patients versus none in the mute group. Postoperative aseptic meningitis occurred only once in the 5 mute patients and in 20 of the 58 nonmute children.

The most significant finding was the postoperative dysarthria, which was found in all our patients after their recovery from mutism, whereas only 3 of 58 children had dysarthria

TABLE 2. Comparison of Characteristics among the Patients with or without Cerebellar Mutism

	Patients with Mutism	Patients without Mutism	Statistical Significance[a]
Number	5	58	
Mean age (mo)	82	83	NS
Sex distribution: M/F	2/3	30/28	NS
Patients with papilledema	4 (80%)	18 (31%)	$P < 0.05$
Tumor diameter (mm)	35	39	NS
Localization			NS
Vermis	3	20	
Vermis and hemisphere	2	17	
Hemisphere	0	13	
Brain stem	0	5	
Brain stem involvement	4 (80%)	18 (31%)	$P < 0.05$
Cranial nerve deficit			
Oculomotor	4 (80%)	25 (43%)	NS
Facial nerve	3 (60%)	9 (15%)	$P = 0.05$
Lower cranial nerves	0	4 (7%)	NS
Aseptic meningitis	1 (20%)	20 (34%)	NS
Dysarthria	5 (100%)	3 (5%)	$P < 0.0001$
Psychological problem	3 (60%)	8 (14%)	$P < 0.05$

[a] **Statistical significance tested with Student's *t* test for continuous variables and with χ^2 test for nominal data.**

after operations not complicated by mutism ($P < 0.0001$). It is noteworthy that preoperative dysarthria, a very rare symptom of childhood posterior fossa tumor, was present in one of the patients with postoperative mutism. Psychological disturbances, characterized by irritability or indifference to the environment, were found in 3 of the 5 children with cerebellar mutism and in 8 of the other 58 ($P < 0.05$).

The Glasgow Outcome Scale (20) score after a mean follow-up period of 3.8 years is shown in *Figure* 2. Even though they recovered from the mutism, the children who presented with the deficit seem to have a more compromised neurological evolution, with no single child becoming totally normal and independent.

DISCUSSION

The role of the cerebellum in the production of speech is still somewhat controversial. According to Turkstra and Bayles (29), there are five processes in the production of normal speech and language: arousal (ascending reticular system); affect and drive (prefrontal and limbic areas); cognition (dominant hemisphere speech areas); initiation, planning, and coordination (five anatomic substrates); and execution (brain stem cranial nerve nuclei). Together with the supplemental motor area, the Broca region, the basal ganglia, and the thalamus, the cerebellum plays a role as one of the five anatomic regions concerned with coordination and planning of speech.

Dysarthria after cerebellar lesions is well known. In a series of 31 patients with cerebellar dysarthria, Lechtenberg and Gilman (21) found that the region most frequently involved was the paravermian superior part of the left hemisphere. More interesting is the question of whether the cerebellum plays a "cognitive role" in speech production. A review on this interesting debate was published recently (6, 15, 19, 22). Three hypotheses can be put forward to explain mutism with cerebellar lesions.

The absence of speech can be unrelated to the cerebellum itself and can be a psychological reaction of the child to the stress of the operation. This childhood stress reaction is well known in psychiatry (3) and is called "elective mutism," a psychogenic condition of short duration in a child who had adequate mastery of language before becoming uncommunicative. Abnormal behavior after posterior fossa operations can occur, and the mutism has been called "verbal isolation" (9). However, we did not find a very significant relationship between behavioral disturbances and mutism in our series. Mutism is also extremely rare after supratentorial operations, which can be presumed to have the same psychological significance to the young child.

In the second hypothesis, favored by most authors, cerebellar mutism is considered to be an extreme form of atactic dysarthria, the complete inability to articulate any sound whatever. It could be compared with the total static and gait ataxia ("astasia-abasia") frequently found after cerebellar hemorrhage.

Third, the absence of speech could be a sign of cognitive dysfunction that occurs when the connections from the neodentate to the Broca area are interrupted (22). Other undoubtedly cognitive disturbances have been found after surgical procedures on the cerebellum in children (2). A strong argument against a predominantly higher function disturbance is the postmutism dysarthria. In a recently described case (11), however, mutism was followed by dysarthria, but also by dysnomia.

Postoperative mutism in children is increasingly being recognized as a common complication after posterior fossa operations (27). We have been able to trace 48 cases in the literature (*Table* 3), including the four cases of Wisoff and Epstein (31), who considered mutism as part of a larger syndrome of pseudobulbar palsy, and the two cases of Fraioli and Guidetti (14). Not all of these cases are described in

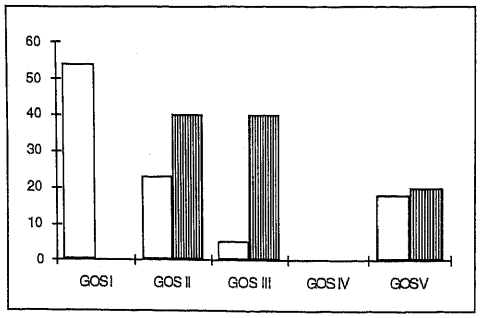

FIGURE 2. Graph illustrates the Glasgow Outcome Scale (*GOS*) score at the moment of latest follow-up in the 58 cases without (*white*) and the 5 cases with (*shaded*) mutism (percentage scale).

TABLE 3. Published Cases with Postoperative Cerebellar Mutism

Series (Ref. No.)	Cases
Fraioli and Guidetti, 1975 (14)	2
Wisoff and Epstein, 1984 (31)	4
Rekate et al., 1985 (26)	6
Yonemasu, 1985 (32)	4
Volcan et al., 1986 (30)	1
Ammirati et al., 1989 (4)	1
Humphreys, 1989 (18)	5
Ferrante et al., 1990 (13)	3
Dietze and Mickle, 1990 (12)	2
Salvati et al., 1991 (28)	1
Nagatani et al., 1991 (24)	1
Herb and Thuyen, 1992 (16)	1
Catsman-Berrevoets et al., 1992 (10)	2
Oiwa et al., 1993 (25)	4
Mooij et al., 1993 (23)	4
Aguiar et al., 1993 (1)	2
Boratynski et al., 1993 (7)	2
Cakir et al., 1994 (8)	1
Asamoto et al., 1994 (5)	1
Crutchfield et al., 1994 (11)	1

sufficient detail to compare them with the present data. Several features, however, emerge as common in all the mutism cases.

The following aspects are similar to our results. When the adult cases of Cakir et al. (8) and of Salvati et al. (28) are excluded, the mean age of the pediatric population is 7.5 years. Seventy-two percent are boys. Sixty-one percent of the cases occurred after the removal of a medulloblastoma. All cases were located in the vermis or in the vermis with extension to one or the other hemisphere. The mutism was transient in every case, after an average interval of 6.5 weeks. In all 38 patients in whom this feature was described, the mutism was followed by dysarthria.

Immediate postoperative mutism was noted in only four cases in the literature. More frequently, an interval (average, 30 h) with more or less normal speech is described. We found this free interval in only one child.

Several questions remain unanswered. The anatomical substrate of the cerebellar lesion in mutism is still not known. Although many cases, both in our series and in the literature, were medulloblastomas, invading the floor of the fourth ventricle, the mutism cannot be explained as a brain stem symptom; these tumors are generally subtotally resected, leaving a fine layer of tumor on the floor of the ventricle, rather than risking a severe brain stem dysfunction. In our population, the occurrence of new brain stem signs did not relate to the mutism.

Most authors suggest the dentate nuclei, located near the midline bilaterally in the roof of the fourth ventricle, to be responsible for the mutism. The major argument is the mutism in the patients described by Fraioli and Guidetti (14), who had small stereotactic lesions in precisely that area. Another argument is the finding that a postoperative CT scan may show bilateral hypodense areas in the dentate nuclei (4). Others (5) have not been able to demonstrate this CT or magnetic resonance imaging abnormality. We have found it very difficult to locate exactly the dentate nuclei on CT scans and to differentiate the postoperative hypodense region bordering the resection bed in cases with or without mutism. No early magnetic resonance imaging studies were done in our patients during the period of mutism.

It seems that the most significant association in our population, postmutism dysarthria, is an important factor in explaining the pathophysiology of the syndrome. Of course, the relationship to these features was already presumed by several authors, but it was never statistically proved in comparison to the children of the same consecutive surgical series who did not suffer from mutism. We conclude, therefore, that our findings corroborate the hypothesis that transient cerebellar mutism is an extreme form of cerebellar (atactic) dysarthria.

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COMMENTS

Although the ideas proposed are not entirely new, the authors have reviewed their own data as well as the published cases of postsurgical mutism. They find that dysarthria occurred in only 5% of posterior fossa surgical patients who did not develop mutism, whereas it occurred in 100% of the children who did develop mutism. They appropriately cite the most recent literature on cerebellar function and support their arguments. This article should stimulate some debate and some further studies of this interesting phenomenon.

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I am surprised at the significant number of cases of cerebellar mutism (5 of 63) that the authors have seen at their institution. I would regard cerebellar mutism after posterior fossa surgery for tumor as an extremely rare event. I personally have never seen a case of cerebellar mutism in my career, although we have had some cases at our institution. I suspect that the lesion responsible for cerebellar mutism is not in the cerebellum but in the floor of the fourth ventricle. I note that one of the patients had a period of coma and stupor of less than 48 hours and a another patient had normal speech before the mutism appeared. In three of the children, the mutism was present directly after surgery. I suspect that there is some transient edema within the floor of the fourth ventricle that leads to the mutism and the subsequent dysarthria.

The authors have made a very strong argument for the mutism being within the cerebellum and particularly within the dentate nucleus. I doubt very much whether this is the case. However, the authors have described the condition of cerebellar mutism well and have pointed out its transient nature. Furthermore, they have stressed the nature of mutism as being a form of severe dysarthria.

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