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The Evaluation and Comparison of Cerebellar Mutism in Children and Adults after Posterior Fossa Surgery: Report of two Adult Cases and Review of the Literature

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Summary

Background. Although there are some cases of cerebellar mutism in adults after posterior fossa surgery for cerebellar tumour it generally occurs in children. Reversible pathophsiology and the anatomical substrate of this syndrome still remain unclear. The predominance of cerebellar mutism in children is suggested to be related to the higher incidence of posterior fossa tumours in children. However, the question regarding the reason for the obvious difference in the incidence of this syndrome between the paediatric and adult population still remaing unanswered. The aim of this study was to evaluate and compare children and adult groups separately to understand the incidence and the clinical characteristics better and to elucidate the pathophysiological basis and predictive factors for this syndrome.

Method. We reviewed, analysed, and compared the cases of cerebellar mutism individually in children and in adults reported in the English literature. We found 106 reported cases in children and 11 cases in adults which were suitable for analysis. We added two adult cases to these.

Findings. The ages of the patients ranged from 2 to 16 (mean, 6.4 year) in children and from 17 to 74 (mean, 38.7 year) in adults. Although vermis was the main location in both groups, the incidence of vermis lesions was considered higher in the paediatric population (%91.5 versus %69.2). The rate of brain stem invasion was prominent in children (%31.1) when compared with adults (%7.6). The latency for the development of mutism and the duration of the mutism were similar in children and adults (mean, 1.4 d versus 2 d and mean, 5.07 wk versus 4.2 wk respectively). Mutism was transient in all the cases of both groups.

Interpretation. Recent concepts of cerebellar physiology disclose the importance of the cerebellum in learning, language, and mental and social functions. Pontine nuclei, the thalamus, motor and sensory areas and supplementary motor areas have been proven necessary for the initiation of speech. It can be hypothesized that uncompleted maturation of the reciprocal links in childhood connecting the cerebellum to these structure makes the children more vulnerable to have postoperative cerebellar mutism in comparison to the adult population.

Keywords: Adult; cerebellar mutism; child; posterior fossa surgery.

Introduction

Mutism is the state of being silent and organic or functional absence of speech in an awake and conscious patient with intact comprehension and no evidence of oral apraxia. Cerebellar damage including degenerative disease, infection, haemorrhage and neoplastic involvement has been known to cause mutism [1, 37]. However, the development of mutism is an uncommon, but well-established syndrome after extensive removal of posterior fossa tumours [4, 22, 29, 30]. Although mutism following stereotactic lesions in the dentate nucleus has been known since 1975 [23], it was first reported by Rekate *et al.* [45] and Yonemasu [60] after posterior fossa surgery in 1985 in separate articles. Although it is well known that the preservation of comprehension and the lack of long-tract lesions and the involvement of the lower cranial nerves is the rule, the pathophysiology of cerebellar mutism has not been elucidated in detail yet. This syndrome must be considered as a separate syndrome from the one described by Wisoff and Epstein [57] in 1984 with presence of long-tract problems and paralysis of lower cranial nerves. Since 1985, a growing number of paediatric patients with cerebellar mutism and subsequent dysarthria have been reported. Although almost all reported cases of mutism after posterior fossa surgery were in children [4, 18, 20, 22, 35, 41] there were a few cases in adults found in the literature [7, 14, 15, 19, 35]. The pathogenesis and the anatomical substrate of this syndrome remain unclear. Although the predominance of cerebellar mutism in children may be attributed to the higher incidence of posterior fossa tumours in

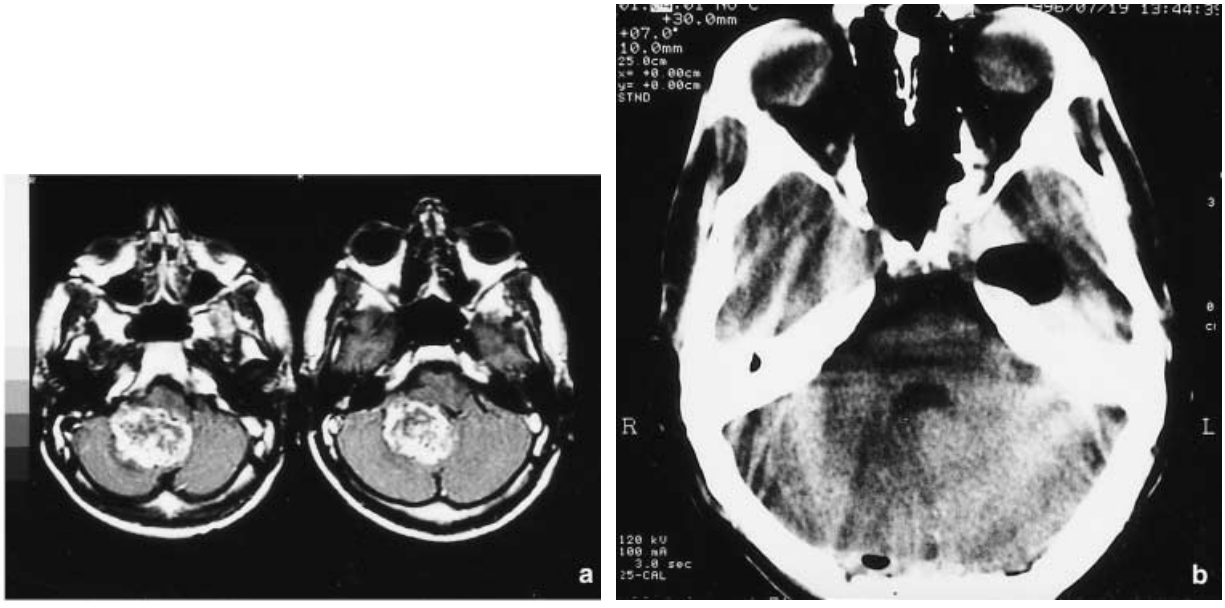

Fig. 1. (a) Axial T_1-weighted magnetic resonance image with gadolinium contrast medium in the right superior vermian area. (b) Computed tomography scan without contrast after the occurrence of the mutism showing no lesion in the area of resection

childhood and to the predilection of vermian tumours, the reason for the noted difference observed in the incidence of this syndrome between the paediatric and adult populations remains unanswered. We reviewed and analysed all the cases of cerebellar mutism after surgery reported in the English literature individually in paediatric and adult patients to understand the incidence, pathophysiology, anatomical substrate, possible predictive factors better for this syndrome.

Case Reports

Case 1

A 32-year-old male patient was admitted to the hospital with complaints of headache nausea, vomiting and ataxia. He displayed an ataxic gate, dysdiadochokinesia and dysmetria in the neurological examination. MR revealed a vermian mass with an extension to the right cerebellar hemisphere (Fig. 1a). An obstructive hydrocephalus was also present. He was operated on in the sitting position. Suboccipital craniectomy was performed. The solid tumour was totally removed through an inferior vermian incision under the operating microscope. The pathological diagnosis was medullablastoma. He was fully alert postoperatively and without additional neurological deficit. The patient became mute on the 2th. postoperative day. Four weeks after the first operation the patient started to utter words. The postoperative CT scan taken following muteness was unremarkable (Fig. 1b). A repeated CT scan one month later showed postoperative changes. The patient was speaking fluently 4 months later.

Case 2

A previously healthy 44-year-old male patient developed progressive ataxia and vomiting. Neurological examination revealed that he had truncal ataxia, bilateral abducens palsies and papillo edema. MRI on admission revealed a posterior fossa mass (Fig. 2a). The patient was operated on and the tumour was totally removed. The pathological diagnosis was astrocytoma. The patient was awake and co-operative without additional neurological deficit in the early postoperative period. On the second day, the patient developed mutism. Early postoperative CT scan following mutism disclosed blood in the tumour bed (Fig. 2b). Three weeks later his speech started to improve gradually but it was still dysarthric until the sixth month postoperatively. In the ninth postoperative month the patient had only mild dysarthria.

Review of the Literature

Paediatric Population

We could find 106 cases of mutism after posterior fossa surgery who were suitable for analysis in the paediatric population (Table 1) [2–5, 7, 8, 11, 12, 14, 17, 18, 20, 22, 25, 30, 32, 34, 36–39, 41, 43, 45, 49, 51, 55–57, 60]. However, we did not include the cases with mutism after brain stem surgery [13, 16, 24] as well as the cases with insufficient data related to the assumed mutism [13, 40, 42, 53, 59] in our review. There were 40 female and 54 male patients. Gender was not mentioned in twelve of the cases [17, 36, 60]. The ages of the patients ranged from 2 to 16 years. Age was not mentioned in Yonemasu's cases [60] as well as in some other studies [8, 12, 36, 39]. Except for these studies, mean age was 6.4 years. The vermis was the main location for the lesions in 97 cases (%91.5). Six of the

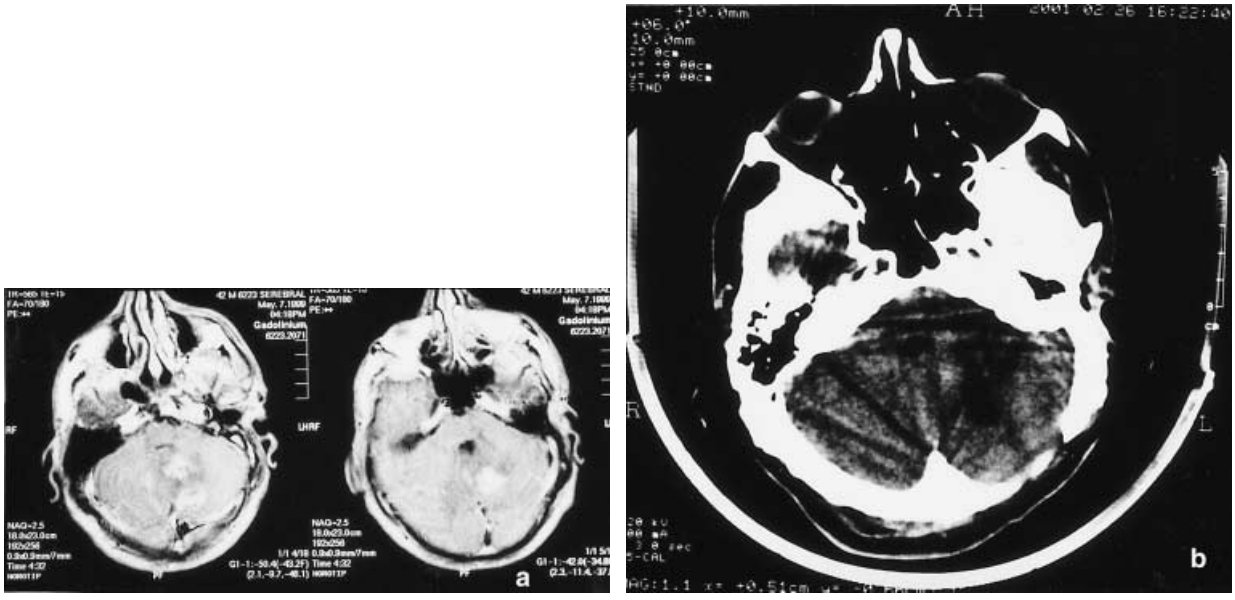

Fig. 2. (a) Axial T_1-weighted magnetic resonance image with gadolinium contrast medium in the left superior vermian area. (b) Computed tomography scan without contrast after the occurrence of the mutism showing blood in the tumour bed

lesions (% 5.6) were located in vermis along with the cerebellar hemisphere. The neoplasm was located in the cerebellar hemisphere in one case [35]. The location of the lesion was not mentioned in two cases [38]. There was brain stem involvement in 33 cases but it was not mentioned in 23 cases. No invasion was reported in 51 cases. The pathological findings of the lesions were as follows: 72 medulloblastomas, 17 astrocytomas, 15 ependymomas, 1 AVM, 1 haematomas. The latency for the development of mutism ranged from 0 to 7 days (mean, 1.4 d). This period was not mentioned for each case in some studies [12, 36, 39]. The mutism lasted from 1 day to 20 wks. (mean, 5.07 wks). This period was also not mentioned for each case in some studies [8, 12, 33, 36]. Hydrocephalus requiring VP shunt developed in 20 cases in the postoperative period. Meningitis developed in 11 cases after surgery. Dysarthric speech developed after mutism in 96 cases. Mutism was transient in all the cases.

Adult Population

A total of 13 cases (over 17 years of age) including two of our own cases (Table 2) [7, 9, 15–17, 19, 35, 50, 57] were reviewed and analysed. There were 9 male and 4 female patients. The age of the patients ranged from 17 to 74 years (mean, 38.7 years). The vermis was the main location in 9 cases. However the other three lesions were located in the cerebellar hemisphere and one of them was located in the fourth ventricle. There was brain stem invasion in one case. Brain stem invasion was not mentioned in two cases. No invasion was reported in ten cases. The pathological findings of the lesions were as follows; 4 medulloblastomas, 3 astrocytomas, 2 metastasis, 2 hemangioblastomas, 1 arterio-venous malformation, 1 choroid plexus papilloma. The latency for the development of mutism ranged from 0 to 6 days (mean, 2 d). The mutism lasted from 4 days to 8 wks (mean, 4.2 wk). This period was not clearly stated in one study [57]. Hydrocephalus requiring a VP shunt and meningitis were not found in this series. Dysarthric speech developed after mutism in 12 cases. We observed that mutism was transient in the paediatric population as well as in the adult population.

Discussion

The term mutism which is an extreme form of atactic dysarthria is usually defined as the complete absence of speech without other aphasic symptomatology in a conscious patient [26, 27]. Although the cerebellum has been implicated in speech output since 1917 by Holmes, [31] its role in speech is still debated. The main speech problems related to the cerebellum including the articulation of syllables, mispronunciation, slow rhythm, absence of stress, distortion of vowels are collected under the term ''dysarthria'' [6,

Table 1. *Analysis of Children with Mutism after Resection of Posterior Fossa Lesions*

	Patient No	Gender	Age	Tumour location	Brain stem invasion	Pathology	Latency of mut.	Duration of mut.	Hydrocephalus postop	Meningitis postop	Dysartria after mut.
Wisoff and Epstein, 1984 [57]	1	M	6	vermis	no	astrocytoma	1	2 wks	no	no	yes
	2	M	3.5	vermis	no	medulloblastoma	3	1 wk	no	no	yes
Rekate *et al.*, 1985 [45]	3	M	6	vermis and cerebellar hemisphere	?	astrocytoma	3	12 wks	no	yes	yes
	4	F	8	vermis	?	medulloblastoma	1	12 wks	no	no	yes
	5	?	2	vermis	?	epandymoma	1	8 wks	no	no	yes
	6	?	9	vermis	?	medulloblastoma	1	12 wks	no	no	yes
	7	?	10	vermis	?	medulloblastoma	1	8 wks	no	no	yes
	8	?	11	vermis	?	medulloblastoma	1	3 wks	no	no	yes
Yonemasu 1985 [60]	9	?	?	vermis	?	ependymoma	1	4 wks	no	no	no
	10	?	?	vermis	?	ependymoma	2	4 wks	no	no	no
	11	?	?	vermis	?	medulloblastoma	2	12 wks	no	no	no
	12	?	?	vermis	?	medulloblastoma	3	12 wks	no	no	no
Volcan *et al.*, 1986 [56]	13	F	8	vermis	no	medulloblastoma	1	2 wks	no	no	yes
Ammirati *et al.*, 1989 [4]	14	F	14	vermis	?	astrocytoma	1	3 wks	yes	no	yes
Humphreys, 1989 [32]	15	M	3	vermis	l lateral wall of the 4th ventricle	medulloblastoma	0	7 wks	yes	yes	no
	16	M	4.5	cerebellar hemisphere	floor and lateral wall of the 4th vent.	medulloblastoma	2	7 wks	no	no	no
	17	M	7	vermis	lateral walls and floor of the 4th ventricle	medulloblastoma	0	10 wks	no	yes	no
	18	M	7	vermis	floor of the 4th ventricle	medulloblastoma	1	16 wks	yes	no	no
	19	F	10	vermis	lateral wall of the 4th ventricle; roof of aqueduct	ependymoma	1	16 wks	yes	no	no
Ferrante *et al.*, 1990 [22]	20	M	6	vermis	r lateral recess of 4th ventricle	astrocytoma	1.5	8 wks	no	yes	yes
	21	F	5.5	vermis	r lateral recess of 4th ventricle	astrocytoma	2	8 wks	no	yes	yes
	22	F	9	vermis	r lateral recess of 4th ventricle	astrocytoma	2	4 wks	no	yes	yes
Dietz and Mickle, 1991 [18]	23	M	7	vermis	the floor of 4th ventricle	medulloblastoma	0	6 wks	no	no	yes
	24	F	15	vermis	no	AVM	0	12 wks	yes	no	yes
Nagatani *et al.*, 1991 [41]	25	F	4	vermis	no	medulloblastoma	1	11 wks	no	no	yes
Gaskill and Marlin 1992 [25]	26	F	8	vermis	r lateral wall of 4th ventricle	medulloblastoma	0	2 wks	no	no	yes

Castman-Berrevoets *et al.*, 1992 [11]	27	M	6	vermis	?	medulloblastoma	1	8.5 wks	no	no	yes
	28	M	8	vermis	?	medulloblastoma	1	8 wks	no	no	yes
	29	F	8	vermis	?	medulloblastoma	2	10 wks	no	no	yes
Herb and Thyen, 1992 [29]	30	M	9	vermis	no	medulloblastoma	2	10 wks	no	no	yes
Al-Jarallah *et al.*, 1994 [3]	31	M	5	vermis	no	astrocytoma	1	3 wks	no	no	yes
	32	F	9	vermis	no	medulloblastoma	1	12 wks	no	no	yes
	33	F	13	vermis	no	medulloblastoma	2	3 wks	yes	no	yes
Asamoto *et al.*, 1994 [5]	34	F	8	vermis	no	medulloblastoma	0.5	2 wks	no	no	yes
van Dongen *et al.*, 1994 [55]	35	M	6	vermis	no	medulloblastoma	1	5 wks	no	no	yes
	36	F	8	vermis	no	medulloblastoma	2	5 wks	no	no	yes
	37	M	8	vermis	the floor of the 4th ventricle	medulloblastoma	1	10 wks	yes	yes	yes
	38	M	5	vermis	no	medulloblastoma	0	8 wks	no	no	yes
	39	M	4	vermis	mesencephalon	ependymoma	1	2 wks	yes	no	yes
Crutchfield *et al.*, 1994 [14]	40	M	7	vermis	?	medulloblastoma	1	7 wks	no	no	yes
Aguiar *et al.*, 1995 [2]	41	M	9	vermis	?	medulloblastoma	2	5 wks	yes	no	no
	42	M	5	vermis	?	medulloblastoma	2	6 wk	no	no	yes
Erşahin *et al.*, 1995 [20]	43	F	7	vermis	no	medulloblastoma	6	3 wks	no	no	yes
	44	M	6	vermis	no	astrocytoma	2	3 wks	yes	no	yes
	45	M	11	vermis	no	medulloblastoma	5	4 wks	no	no	yes
	46	F	11	vermis	no	medulloblastoma	5	2 wks	yes	no	yes
	47	M	11	vermis	no	medulloblastoma	2	1.5 wks	no	yes	yes
	48	M	3	vermis	no	medulloblastoma	3	4 wks	no	no	yes
	49	M	3.5	vermis	no	medulloblastoma	5	6 wks	no	no	yes
Pollack *et al.*, 1995 [43]	50	F	6	vermis	4th ventricle floor and r lateral recess	medulloblastoma	2	6 wks	no	no	yes
	51	F	11	vermis	l lateral recess of the 4th ventricle	ependymoma	1	2 wks	no	no	yes
	52	M	9	vermis	no	medulloblastoma	1	4 wks	no	no	yes
	53	M	10	vermis	4th ventricle floor	medulloblalstoma	0	3 wks	no	no	yes
	54	M	16	vermis	no	astrocytoma	2	4 wks	no	no	yes
	55	M	9	vermis	4th ventircle floor	medulloblastoma	0	4 wks	no	no	yes
	56	F	6	vermis	l middle cerebellar peduncle	astrocytoma	3	2 wks	no	no	yes
	57	M	9	vermis	both middle cerebellar peduncle	ependymoma	2	2 wks	no	no	yes
	58	F	8	vermis	l 4th ventricle floor	medulloblastoma	3	1 wk	no	no	yes
	59	M	5	vermis	r floor of the 4th	medulloblastoma	3	2 wks	no	no	
	60	M	3.5	vermis	floor of the 4th ventricle	astrocytoma	3	2 wks	no	no	yes
	61	M	4	vermis with cerebellar hemispheres	4th ventricle floor	medulloblastoma	3	2 wks	no	no	yes

Table 1. *Continued*

	Patient No	Gender	Age	Tumour location	Brain stem invasion	Pathology	Latency of mut.	Duration of mut.	Hydrocephalus postop	Meningitis postop	Dysartria after mut.
Calenberg *et al*, 1995 [8]	62–66	3F, 2M	41–152 months	3 vermis 2 vermis and hemisphere	4 brain stem involvement	3 medulloblastoma 1 astrocytoma 1 ependymoma	4 cases 1 case 7	2–11 wks	yes one case	on	yes
Dailey *et al.*, 1995 [17]	67	M	4	vermis	brain stem involvement	ependymoma	1	2 wks	no	no	yes
	68	F	12	vermis	no	medulloblastoma	0	1 wks	no	no	yes
	69	F	9	vermis	brain stem involvement	medulloblastoma	$\frac{1}{2}$	4 wks	no	no	yes
	70	F	6	vermis	no	medulloblastoma	$\frac{1}{2}$	12 wks	no	no	yes
	71	M	3	vermis	brain stem involvement	medulloblastoma	1	$\frac{1}{2}$ wks	no	no	yes
	72	F	6	vermis	no	astrocytoma	1	6 wks	no	no	yes
	73	M	2,5	vermis	brain stem involvement	medulloblastoma	2	3 wks	no	no	yes
	74	F	10	vermis	no	medulloblastoma	3	3 wks	no	no	yes
Salvati *et al.*, 1996 [51]	75	M	11	vermis with cerebellar hemisphere	?	medulloblastoma	1	2 wks	no	no	yes
	76	M	5	vermis with cerebellar hemisphere	?	medulloblastoma	3	2 wks	no	no	yes
Mastrionardi, 1996 [39]	77	F	3	vermis	?	ependymoma	0	3 wks	no	no	yes
Jones, 1996 [34]	78	F	10	vermis	no	medulloblastoma	2	12 wks	no	yes	yes
Bhatoe, 1997 [7]	79	F	4	vermis	?	ependymoma	0	1 wk	no	no	yes
Koh *et al.*, 1997 [36]	80–83	?	6–12	vermis	no	4 medulloblastoma	1–5	2–8 wks	no	3 yes	yes
Liu *et al.*, 1998 [38]	84	F	7	vermis	brain stem involvement	medulloblastoma	0	4 wks	no	no	yes
	85	M	3.5	not stated	?	medulloblastoma	0	3 wks	no	no	yes
	86	F	3.5	not stated	?	medulloblastoma	0	2 wks	yes	no	yes
	87	M	7	vermis	brain stem involvement	ependymoma	0	3 wks	yes	no	yes
Sinha, 1998 [53]	88	M	7	vermis	no	hematoma	1	3 wks	no	no	yes
JanBen, 1998 [33]	89–94	3F 3M	4–10	vermis	no	3 medulloblastoma 2 astrocytoma 1 ependymoma	2–6	3 wks–4 ms	yes 5	no	yes
Catsman-Berrevoets *et al.*, 1999 [12]	95–106	8M 4F	2–16	vermis	no	8 medulloblastoma 1 ependymoma 2 P1. astrocytoma	?	1 d–5 ms	no	no	yes

Table 2. *Analysis of the Adults with Mutism after Resection of Posterior Fossa Lesions*

	Patient No	Gender	Age	Tumor location	Brain stem invasion	Pathology	Latency of mut.	Duration of mut.	Hydrocephalus postop	Meningitis postop	Dysartria after mut.
Wisoff and Epstein, 1984 [57]	1	M	17	vermis	l lateral recess of the 4th ventricle	astrocytoma	2	sev. wks	no	no	yes
Salvati *et al.*, 1991 [50]	2	M	20	vermis	no	medulloblastoma	2	4 wks	no	no	yes
D'Avanzo *et al.*, 1993 [16]	3	M	45	vermis	not stated	medulloblastoma	2.5	8 wks	no	no	yes
	4	F	20	vermis	not stated	medulloblastoma	3	6 wks	no	no	yes
Çakir *et al.*, 1995 [15]	5	M	61	cerebellar hemisphere	no	metastasis	0	4 d	no	no	no
Dailey *et al.*, 1995 [17]	6	F	20	vermis	no	astrocytoma	12 hrs	8 wks	no	no	yes
Bhatoe, 1997 [7]	7	M	28	cerebellar hemisphere	no	hemangioblastoma	2	4 wks	no	no	yes
Dunwoody *et al.*, 1997 [19]	8	M	54	vermis	no	AVM	0	3 wks	no	no	yes
Kai *et al.*, 1997 [35]	9	M	71	hemisphere	no	hemangioblastoma	2	4 wks	no	no	yes
	10	F	74	vermis	no	metastasis	2	3 wks	no	no	yes
Caner *et al.*, 1999 [9]	11	F	18	4th ventricle	no	choroid plexus pap.	6	4 wks	no	no	yes
İldan *et al.*, 2001	12	M	32	vermis	no	medulloblastoma	2	4 wks	no	no	yes
	13	M	44	vermis	no	astrocytoma	2	3 wks	no	no	yes

37, 45]. A new form of mutism was added to these types by Rekate *et al.* [45] in 1985 termed as "cerebellar mutism". A growing number of paediatric patients with mutism following posterior fossa surgery have been published since the description of this syndrome by Rekate *et al.* [45]. Although nearly all the cases of mutism following posterior cranial fossa surgery occurred in children there were also a few young adult or adult cases mentioned in the literature.

The anatomical location of the cerebellar lesion in mutism and formation theories remain controversial. Two main theories have been suggested on what contributes to the occurrence of mutism [22]. The first theory is related to the psychological background of cerebellar mutism. The absence of speech can be a psychological reaction of the child to the stress of the operation. The childhood stress reaction termed as 'elective mutism' is a well-known psychogenic condition which includes behavioural changes, and refusal of food intake in addition to mutism. The psychological hypothesis suggests that the child produces a sense of negativity when he/she feels betrayed by his parents or the doctors and reflects it with elective mutism. The most important characteristic of this type of mutism is that there is recovery of verbal expression without a dysarthric period as soon as the child goes back home after the hospital stay [35]. Although psychological factors have been proposed as causative factors, there are very few patients who showed recovery from their mute state upon returning home [20, 22, 35]. Psychological interpretation cannot explain why mutism occurs only in children who undergo operations for posterior fossa lesions and why the speech recovery phase is commonly prolonged by dysarthria. The same psychological significance should have been a causative factor after supratentorial operations too. However, mutism is extremely rare in such cases. It is highly possible that psychological stress and prolonged hospitalisation are considerable secondary factors retarding the recovery of the faculty of speech after this syndrome.

The second main theory is related to organic background of cerebellar mutism. However the causes, pathophysiology and anatomical basis for mutism in children and adults with posterior fossa tumour have remained conjectural up to date. Numerous attempts have been made to explain the pathophysiology and anatomical basis of cerebellar mutism.

Most authors reported the damage located in the cerebellar peduncles, vermian or paravermian zone

involving dentate nuclei, and hemispheric areas to be responsible for mutism [18, 23, 35, 45]. Dietze and Mickle [18] suggested that injury in the midportion of the cerebellum with or without dentate nuclei involvement increases the risk of occurrence of postoperative cerebellar mutism. Fraioli and Guidetti [23] presented two cases of mutism after bilateral stereotactic lesions of dentate nuclei and the interpositus nucleus. These nuclei send efferents to the paravermian portion considered to be the area of cerebellum that controls phonation [54]. Ammirati *et al.* [4] suggested another argument about the role of the dentate nuclei in cerebellar mutism by detecting bilateral hypodense areas in dentate nuclei. But this observation on CT could not be confirmed by other authors [5, 20].

Siegfried *et al.* [52] suggested that mutism occurred after bilateral thalamotomy for Parkinson' disease. Ricklan *et al.* [46, 47] likewise reported some communication problems involving articulation and phonation in the immediate postoperative period in patients who had undergone bilateral ventrolateral thalamotomy. This argument possibly indicates the contribution of the superior cerebellar peduncles which connect the dentate nuclei in a crossed fashion to the contralateral red nucleus and thalamus. On the other hand, Crutchfield *et al.* [14] suggested that bilateral interruption of the dentatothalamocortical pathways might be responsible for the occurrence of postoperative mutism. Further evidence supporting and confirming this theory has been published by Frim and Ogilvy [24]. They stated that interruption or disturbance of the dentatorubral and dentothalamocortical tracts may produce the same clinical syndrome seen after direct injury to the cerebellar hemisphere or median structures after posterior fossa operation. The possibility of dysfunction of the ascending mesencephalofrontal fibers originating from A9 and A10 dopaminergic cell groups was suggested as another theory [10].

Pollack *et al.* [43] also reported pseudobulber signs and emotional changes and/or decreased initiation of voluntary movements after posterior fossa surgery and concluded that inferior vermian incisions might be responsible for the cerebellar mutism in midline cerebellar mass lesions.

Postoperative cerebellar mutism can be seen almost exclusively with midline cerebellar mass lesions [1, 37]. We found that the lesions were located in the vermis with or without extension to one or other hemisphere in the majority of the reported cases of mutism in children (%91.5) and adults (%69.2). However, the tumour was located in the right hemisphere in the patient reported by Çakir *et al.* [15]. Lechtenberg and Gilman [37] suggested that speech function was located in the right cerebellar hemisphere in left handed people and vice versa for the right handed people and they claimed that the dominant hemisphere lesion was responsible for the cerebellar mutism.

The common posterior fossa tumours have all been implicated, including medulloblastoma (67.9%), astrocytomas (16.0%), ependymomas (14.1%), AVM (0.9%), haematomas (0.9%) in children and medulloblastoma (30.7%), astrocytomas (23.0%), metastasis (15.3%), hemangioblastomas (15.3%), arterio-venous malformation (%7.6), choroid plexus papillomas (7.6%) in adults. Thus, this syndrome does not seem tumour specific as stated by Humphreys [32].

Some authors have stressed the importance of postoperative hydrocephalus or meningitis on the pathophysiology of cerebellar mutism. Although the high incidence of postoperative hydrocephalus and meningitis were also emphasized by Humphreys [32] and Ferrante *et al.* [22] in their series, Gonzalez and Villa [28] concluded that there was no relationship between hydrocephalus, meningitis and cerebellar mutism in their extensive review of the literature. We found that hydrocephalus requiring VP shunting developed in 18.8% of the cases and meningitis in 10.3% of the cases in the postoperative period in children. On the other hand hydrocephalus requiring VP shunting and meningitis were not detected in adults. Although disturbance of cerebrospinal fluid circulation and postoperative meningeal reactions were considered as possible precipitating factors in some studies [21], these complications may simply be co-incidental since most of the patients in children and none of the patients in adults had neither hydrocephalus nor meningitis.

Immediate postoperative mutism was noted in only 21 paediatric cases [6] (17.9%) and 2 (15.3%) in adult cases. We found that mutism usually develops several days after surgery (mean, 1.4 d in children, 2 d in adults). The presence of this interval indicates that the structures responsible for this syndrome do not suffer from direct injury during surgery. The finding of an interval of normal verbal expression which is present in the majority of cases, suggests that other factors such as tissue swelling or ischemia due to spasm of one or more vessels supplying the nuclei or other cerebellar structures may trigger the prime cerebellar disturbance. Ferrante *et al.* [22] suggested that spasm of one or more vessels supplying the nuclei or other cerebellar struc-

tures and/or vasogenic edema of this area might precipitate the cerebellar mutism. This is analogous to the clinical picture of akinetic mutism arising as a result of occlusion of a perforating branch of the mesencephalic artery.

The duration of the mutism was 5.07 wks in children, 4.2 wks in adults. After the resolution of mutism, the children with mutism suffered from dysarthric speech lasting for months after the surgical intervention. Dysarthria followed mutism in almost all the cases in the literature. We found that in ten of the paediatric patients (9.4%) and in one adult patient (7.6%), dysarthric speech was not detected after resolution of the mutism.

It is conceivable that the age distribution of the patients in cerebellar mutism is related to the higher incidence of posterior fossa tumours and to the predilection for vermian tumours in childhood. However a recent report by Riva and Giorgi [48] stated that the cerebellum is important not only for motor execution but also for the planning and initiation of learned mechanisms as a modulator of mental and social functions and this role is also operative in early childhood. The reciprocal links connecting the cerebellum to the structures including pontine nuclei, thalamus, motor and sensory areas and supplementary motor areas that have been proved necessary for the initiation of speech and mental and social functions are operative but vulnerable to edema, ischemia and surgical trauma in childhood in comparison to adults since myelination proceeds rapidly within the brain up to about 24 months of age. This process slows down markedly thereafter, although fibers in the associating areas of the brain continue to myelinate through the first several decades of life [58]. If cerebellar mutism observed after posterior fossa surgery can be interpreted as an effect of a major diaschisis of these reciprocal connections, it can be hypothesized that uncompleted maturation of these links in childhood makes children more vulnerable to post operative cerebellar mutism.

Our detailed review of the literature showed that a complex mechanism is responsible for cerebellar mutism. Therefore simultaneous analysis of several factors acknowledge the importance of the interactions of these factors. This syndrome appears to be multifactorial in aetiology.

Cerebellar mutism seems to be more common than previously assumed. Catsman-Berrevoets *et al.* [12] found a higher incidence of this syndrome (29.0%) than reported in the literature (8.0%–13.0%) [17, 43, 44]. Therefore the possibility of developing postoperative mutism should be taken into consideration in the presence of a large cerebellar tumour, particularly in eloquent areas of the cerebellum including midline and paravermian regions. The complication of a postoperative mutism syndrome should be discussed with the patient's family in the pre-operative period and in addition since this syndrome is of a transient nature, necessary adjuvant therapies including chemotherapy and radiotherapy should not be postponed.

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Comment

The aim of this paper is to discuss the possible anatomical and pathogenic mechanisms underlying the appearance of cerebellar

mutism after surgery in the posterior fossa and to investigate factors accounting for the different rate of this event in the paediatric snd in the adult age. The authors present two personal adult cases and review the current English literature on the topic. Overall one hundred and nineteen cases have been analysed, 106 children and 13 adults respectively. Most of them presented lesions located in the vermis (91.5% in the paediatric group and 69.2% in adult patients). The authors speculate that the higher incidence of cerebellar mutism in children is related to the higher frequency of posterior fossa tumours and to the predilection of vermian localization in childhood. Although the aetiology of this clinical event has to be supposed as multifactorial, the likely incomplete maturation of the pathways connecting the cerebellar structures with subcortical and cortical systems responsible of the initiation of speech can provide further support to the prevalence of the phenomenon in the paediatric age.

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