

Inconsistent terminology for cerebellar mutism

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Dear Editor:

We would like to thank Gudrunardottir and coworkers for commenting on our paper “Fronto-cerebellar fiber tractography in pediatric patients following posterior fossa tumor surgery” [1]. We admit that the paper entitled “Cerebellar mutism: review of the literature” by Gudrunardottir et al. [2] should have been cited at the respective passage instead of or even together with “Cerebellar mutism: definitions, classification and grading of symptoms” by the same group [3] which were published both in 2011.

The issue raised by Gudrunardottir et al. is the ambiguous use of the term of “cerebellar mutism (CM)” and its exact definition. In our report, we used CM and CM syndrome (CMS) as synonyms and thereby may not have clearly enough expressed that we agree that posterior fossa syndrome (PFS) describes a broader symptomatology complex of which CMS or CM is an integral part. In this context and to better elucidate the problem of inconsistent terminology, we scrutinized the terminology used in the literature in the period between 2000 and 2013 by performing a Medline search using the phrases “cerebellar mutism” and “cerebellar mutism syndrome.” We excluded all non-English papers and all publications, which did not primarily deal with cerebellar pathology. The search revealed 86 publications: 12 reviews, 13 letters to the editors, 26 case reports, and 35 original papers. Among these, we evaluated the terminology used for the clinical picture dealing with mutism caused by cerebellar pathology. Eventually, we found up to nine different specifications (Table 1). The majority of papers used only one or two terms ($n=57$) for the symptomatology. The mean number of terms used was 2.2

ranging from one up to five terms per paper. CM and CMS were used most often as synonyms ($n=14$). Even more confusing synonymous descriptions were CM together with “posterior fossa syndrome” ($n=5$) or together with “akinetic mutism” ($n=4$). In contrast, clear delineation between the terms CM and CMS was performed in only seven publications.

This wide spectrum of terms employed clearly shows that the definition of CM and its associated syndromes is still without clear definition. Thus, we support the need for a clear and widely accepted terminology proposed by Gudrunardottir et al. [3]. We agree with their definition of CM as a muteness following lesion of the cerebellum as opposed to occurrence with lesions of the cerebrum or the lower cranial nerves. We also agree that CM is only one symptom of the CMS complex that also includes ataxia, hypotonia, and irritability or that CM and CMS are part of PFS that additionally includes cranial nerve deficits, neurobehavioral changes, and urinary retention or incontinence [3]. In our view for future discussions, three issues need further clarification: firstly, any symptoms of decreased impetus may be better classified within CMS rather than in the context of PFS as proposed by Gudrunardottir et al. [3]. This includes impaired impulsion of motor movements (as long as no brainstem lesion is present) or impaired impulsion of eye opening, which may be similar in its cerebellar origin like impaired impulsion of speech production. Secondly, neurobehavioral changes like apathy and deficits of attention or memory may resemble lack of cerebellar control on higher prefrontal function, which is yet not fully understood. As long as anatomical correlates are not clearly identified, an exact and sustainable terminology cannot be found. Nevertheless, it seems reasonable to distinguish CMS with neurological deficits being related to cerebellar functions from PFS that includes symptoms related to cranial nerves or brainstem deficits. Thirdly, grading of severity of CMS is still ill defined. Robertson et al. used the duration of symptoms as a measure for severity [4]. The future goal may be to grade the severity of muteness itself as well as to grade the amount of symptoms within CMS. This may be relevant to evaluate even mild

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Table 1 Review of the syndrome terminology mutism symptoms caused by cerebellar pathology in the available literature from 2000 to 2013 ($n=86$) and their use for differentiation or as synonyms

Terminology	Cited (n)	Used for differentiation (n)	Used as synonyms (n)
Cerebellar mutism (CM)	77	CM vs. CMS	7
Cerebellar mutism syndrome (CMS)	27		CM and PFS
Posterior fossa syndrome (PFS)	35	PFS = CM+	15
Transient cerebellar mutism (tCM)	14		CM and MSD
Mutism with subsequent dysarthria (MSD)	9		CM and CCAS
Akinetic mutism (akM)	8	akM in frontal lesions	5
Cerebellar cognitive affective syndrome (CCAS)	16	CCAS term for adults	10
Transient cerebellar eye closure (TCES)	3		PFS and CCAS
Cerebellar syndrome (CS)	4		PFS and akM
			CCAS and akM

CM+ describes cerebellar mutism together with symptoms like hypotonia, ataxia, neurobehavioral changes, and emotional lability

symptomatology that occurs most probably more often than reported. In conclusion, we agree that there is the need of an interdisciplinary consensus, which allows comparability of clinical research in this field.

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