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We read the article of Miller et al[1] with great interest. In this retrospective study, the neuroimaging data of a group of 11 children with postoperative posterior fossa syndrome (PFS) and an age- and gender-matched control population who did not develop the syndrome were analyzed to evaluate whether pediatric patients with postoperative PFS present with a consistent pattern of surgical damage and perfusion alterations at the supratentorial level as reflected by cerebellocerebral diaschisis (CCD). A consistent pattern of bilateral structural damage to the proximal efferent cerebellar pathway (pECP) was found in association with a significant decrease of cerebral blood flow, mainly within the frontal brain regions. Based on these findings, the authors concluded that bilateral surgical damage to the pECP is the primary cause of PFS and that predominantly frontal CCD acts as the underlying pathophysiologic mechanism.

We fully agree that the study of Miller et al[1] adds important evidence to current insights in the possible cause and pathophysiologic mechanism underlying the PFS, but we would like to point out that CCD has already been reported on several previous occasions as a possible explanation for transient postoperative mutism[2,3] and for the constellation of neurobehavioral and affective symptoms characterizing PFS in the pediatric population.[4-6] For instance, Mariën et al (2001, 2003)[4,6] reported the preliminary results of a prospective study in which the pathophysiologic role of CCD in PFS was explored. An illustrative patient was described who, in the preoperative phase, already presented with mild dysexecutive symptoms reflected on technetium 99m hexamethylpropyleneamine oxime single-photon emission CT (SPECT) by perfusional changes in the anatomo-clinically suspected prefrontal brain regions. After surgical resection of a posterior fossa medulloblastoma, full-blown PFS was associated with a significant aggravation and extension of the preoperative supratentorial perfusional deficits on repeat SPECT. When akinetic mutism receded and behavioral and affective distortions started to ameliorate after a 5-week period, a marked improvement of regional cerebral blood flow was objectified bilaterally in the prefrontal areas. By sharp contrast, no pre- and postoperative supratentorial perfusion alterations were observed in a child who did not develop PFS after posterior fossa tumor resection.

On the basis of the close parallelism between the development and course of neurobehavioral symptoms and perfusional changes on SPECT in the anatomo-clinically suspected supratentorial brain regions, we concluded that CCD might be intrinsically implicated in the pathophysiology of the PFS. As consistently reflected by CCD, the distant metabolic impact of surgical damage to the cerebellum via the dentatorubrothalamic tract on the supratentorial brain regions crucially involved in language dynamics and behavioral and affective regulation was later confirmed in an adolescent patient as well as in a larger study of 5 children with PFS.[7,8]

The authors infer from the correlation between PFS symptoms and perfusional changes within the frontal lobe regions that "mutism in PFS is probably a speech apraxia rather than a simple dysarthria, dysphasia, or other cerebellar speech disorder."[1] Although we are strongly inclined to support the view that patients with PFS, in addition to subsequent cerebellar dysarthria, may also present with higher level nonmotor speech and language disorders such as apraxia of speech or even aphasia, there still are no studies that substantiate the view of the apractogenic nature of the verbal output disorder following the phase of postoperative mutism.[9] Given the fact that apraxic speech symptoms have already been described as CCD phenomena in adult patients after focal cerebellar damage, more systematic studies are needed to elucidate the exact nature and pathophysiologic substrate of speech and language phenomena following the phase of mutism in PFS.[10,11]

References

1. Miller NG, Reddick WE, Kocak M, et al. **Cerebellocerebral diaschisis is the likely mechanism of postsurgical posterior fossa syndrome in pediatric patients with midline cerebellar tumors.** *ANJR Am J Neuroradiol* 2010;31:288–94. Epub 2009 Oct 1
2. Germano A, Baldari S, Caruso G, et al. **Reversible cerebral perfusion alterations in children with transient mutism after posterior fossa surgery.** *Childs Nerv Syst* 1998;14:114–19
3. Sagiuchi T, Ishii K, Aoki Y, et al. **Bilateral crossed cerebello-cerebral diaschisis and mutism after surgery for cerebellar medulloblastoma**. *Ann Nucl Med* 2001;15:157–60
4. Mariën P, Engelborghs S, Fabbro F, et al. **The lateralized linguistic cerebellum: a review and new hypothesis.** *Brain Lang* 2001;79:580–600
5. Clerico A, Sordi A, Ragni G, et al. **Brief report: transient mutism following posterior fossa surgery studied by single photon emission computed tomography (SPECT).** *Med Pediatr Oncol* 2002;38:445–48
6. Mariën P, Engelborghs S, Michiels E, et al. **Cognitive and linguistic disturbances in the posterior fossa syndrome in children: a diaschisis phenomenon?** *Brain Lang* 2003;87:162
7. Baillieux H, De Smet HJ, Lesage G, et al. **Neurobehavioral alterations in an adolescent following posterior fossa tumor resection**. *Cerebellum* 2006;5:289–95
8. De Smet HJ, Baillieux H, Wackenier P, et al. **Long-term cognitive deficits following posterior fossa tumor resection: a neuropsychological and functional neuroimaging follow-up study.** *Neuropsychology* 2009;23:694–704
9. Riva D, Giorgi C. **The cerebellum contributes to higher cognitive functions during development: evidence from a series of children surgically treated for posterior fossa tumors.** *Brain* 2000;123:1051–61
10. Mariën P, Verhoeven J, Engelborghs S, et al. **A role for the cerebellum in motor speech planning? Evidence from a case with the foreign accent syndrome.** *Clin Neurol Neurosurg* 2006;108:518–22. Epub 2005 Aug 1
11. Cohen DA, Kurowski K, Steven MS, et al. **Paradoxical facilitation: the resolution of foreign accent syndrome after cerebellar stroke.** *Neurology* 2009;73:566–67

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