Cerebellar mutism prevention

Cerebellar mutism can occur in a third of children undergoing cerebellar resections.

The median suboccipital keyhole telovelar approach provides relative wide access to resect most fourth ventricle tumors completely and with satisfactory results [1].

Development of intraoperative neuromonitoring of the cerebellar efferent pathways as well as improved preoperative risk stratification could help to establish a patient-specific strategy with optimal balance between the degree of resection and functional integrity [2].

The wide dissection of the cerebellomedullary fissure and gentle tonsils retraction may prevent the occurrence of cerebellar mutism or other major cerebellar dysfunctions [3].

Recent evidence proposes it may arise from uni- or bilateral damage of cerebellar efferents to the cortex along the dentatothalamic tract.

There are no certain ways to prevent the post-operative pediatric CMS from developing. Pre-operative corticosteroids have been recommended to reduce peritumoral edema, which may decrease nausea and vomiting and improve appetite and neurological symptoms. Intra-operative electrophysiological monitoring of the lateral rectus and facial muscles may help when the surgeon is working near the 6th cranial nerve [4], and surgical access and technique seem to play an important role. Electrophysiological monitoring, the avoiding of vermian dissection in favor of a telovelar approach, as well as minimal use of retraction and ultrasonic aspiration at the Children's National Medical Center in Washington decreased the incidence of the syndrome from 39% to 13% over an 8 year period [5] another institute similarly reported lower incidence after abandoning vermal split

---

Properly designed multicenter trials are needed to provide stronger evidence regarding effective prevention of cerebellar mutism and the best therapeutic approaches for such patients with a combination of pharmacological agents and multidisciplinary speech and behavior augmentation [6].

At present, no neurophysiological procedure is available to monitor this pathway intraoperatively. Giampiccolo et al. specifically aimed at filling this gap.

They assessed 10 patients undergoing posterior fossa surgery using a conditioning-test stimulus paradigm. Electrical conditioning stimuli (cStim) were delivered to the exposed cerebellar cortex at interstimulus intervals (ISIs) of 8-24 ms prior to transcranial electric stimulation of the motor cortex, which served as test stimulus (tStim). The variation of motor-evoked potentials (MEP) to cStim + tStim compared with tStim alone was taken as a measure of cerebello-cortical connectivity.

cStim alone did not produce any MEP. cStim preceding tStim produced a significant inhibition at 8 ms ($p < 0.0001$) compared with other ISIs when applied to the lobules IV-V-VI in the anterior cerebellum and the lobule VIIB in the posterior cerebellum. Mixed-effects of decrease and increase in MEP amplitude were observed in these areas for longer ISIs.

The inhibition exerted by cStim at 8 ms on the motor cortex excitability is likely to be the product of activity along the cerebello-dento-thalamo-cortical pathway. They showed that monitoring efferent cerebellar pathways to the motor cortex is feasible in intraoperative settings. This study has promising implications for pediatric posterior fossa surgery with the aim to preserve the cerebello-cortical pathways and thus prevent cerebellar mutism [7].

References

1)
Wang J, Wang X, Xu J, Wu Z, Dou Y. Microsurgical Management of Fourth Ventricle Lesions Via the Median Suboccipital Keyhole Telovelar Approach. J Craniofac Surg. 2022 Aug 15. doi: 10.1097/SCS.0000000000008883. Epub ahead of print. PMID: 35968951.

2)
Beez T, Munoz-Bendix C, Mijderwijk HJ, Remke M, Hänggi D. Structural damage burden and hypertrophic olivary degeneration in pediatric postoperative cerebellar mutism syndrome. Neurosurg Rev. 2022 Aug;45(4):2757-2765. doi: 10.1007/s10143-022-01791-7. Epub 2022 Apr 20. PMID: 35441993; PMCID: PMC9349162.

3)
Tomasello F, Conti A, Angileri FF, Cardali S. Telo-velar approach to fourth-ventricle tumors: how I do it. Acta Neurochir (Wien). 2015 Apr;157(4):607-10. doi: 10.1007/s00701-015-2358-z. Epub 2015 Feb 6. PubMed PMID: 25652723.

4)
Albright AL, Pollack IF, & Adelson PD (2008). Principles and Practice of Pedatric Neurosurgery (Second Edition) New York: Thieme

5)
Siu A, Wells EM, Snyder K, Defreitas T, Myseros JS, Yaun AL, Magge S, Oluigbo C, Keating RF (2013) Cerebellar Mutism Syndrome: A significant reduction in incidence and risk factor analysis in children with medulloblastoma. Presented at International Society of Pediatric Neurosurgery Annual Meeting, Mainz, Germany, September 30, 2013, publication in prep. Personal Communication

6)
Pitsika M, Tsitouras V. Cerebellar mutism. J Neurosurg Pediatr. 2013 Dec;12(6):604-14. doi: 10.3171/2013.8.PEDS13168. Epub 2013 Sep 27. Review. PubMed PMID: 24073751.

7)
Giampiccolo D, Basaldella F, Badari A, Squintani GM, Cattaneo L, Sala F. Feasibility of cerebello-cortical stimulation for intraoperative neurophysiological monitoring of cerebellar mutism. Childs Nerv Syst. 2021 Apr 9. doi: 10.1007/s00381-021-05126-7. Epub ahead of print. PMID: 33835202.