ABSTRACT

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Cerebellar Mutism Syndrome in Pediatric Neuro-oncology: A Multidisciplinary Perspective and Call for Research Priorities.

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Cerebellar mutism syndrome (CMS), also known as posterior fossa syndrome, occurs in a subset of children after posterior fossa tumor resection, most commonly medulloblastoma. Patients with this syndrome exhibit often transient, although protracted, symptoms of language impairment, emotional lability, cerebellar, and brainstem dysfunction. However, many patients experience persistent neurological deficits and lasting neurocognitive impairment. Historically, research and clinical care were hindered by inconsistent nomenclature, poorly defined diagnostic criteria, and uncertainty surrounding risk factors and etiology. Proposed diagnostic criteria include two major symptoms, language impairment and emotional lability, as proposed by the international Board of the Posterior Fossa Society in their consensus statement as well as other experts in this field. Risk factors most commonly associated with development of CMS include midline tumor location, diagnosis of medulloblastoma and specific tumor subtype, younger age at diagnosis, and preoperative language impairment. A proposed etiology of CMS includes disruption of the cerebellar outflow tracts, the cerebellar nuclei, and their efferent projections through the superior cerebellar peduncle. Treatment for CMS remains supportive. Herein, we present a comprehensive overview of CMS etiology, diagnosis, risk factors, clinical presentation, and clinical management. In addition, we identify essential multidisciplinary research priorities to advance diagnostics, prevention, and intervention efforts for patients with, or at risk for, development of CMS.

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