Cerebellar mutism syndrome

- Cerebellar Mutism/Posterior Fossa Syndrome Following Resection of Posterior Fossa Tumor in Pediatric Patients: Assessing Pathophysiology, Risk Factors, and Neuroradiographic Features
- Therapeutic potential of acetyl-DL-leucine and its L-enantiomer in posterior fossa syndrome: Mechanistic insights
- The Current State of Research on Postoperative Cerebellar Mutism Syndrome: A Bibliometric Analysis
- Tumour volume as a predictor of postoperative speech impairment in children undergoing resection of posterior fossa tumours: a prospective, multicentre study
- Radiological Predictors of Cognitive Impairment in Paediatric Brain Tumours Using Multiparametric Magnetic Resonance Imaging: A Review of Current Practice, Challenges and Future Directions
- Postoperative word-finding difficulties in children with posterior fossa tumours: a crosslinguistic European cohort study
- Infradentate Approach to the Fourth Ventricle with Tubular Retraction System for Medulloblastoma: Feasibility of a Minimally Invasive Technique to Avoid Anatomical Complications in a Pediatric Patient
- Medical management of cerebellar mutism syndrome at a quaternary children's hospital

Cerebellar mutism (CM) was first described by Rekate et al. in 1985 following posterior fossa surgery in children [1] ;since then, it has increasingly been reported, mainly occurring as a postoperative complication.

Posterior fossa syndrome (PFS) and cerebellar mutism are often used interchangeably in the literature.

Epidemiology

Incidence of cerebellar mutism: 11–29% of children following surgery for cerebellar tumors [2] including cerebellar medulloblastoma (53%), posterior fossa ependymoma (33%) & cerebellar pilocytic astrocytoma (11%) [3].

Etiology

It has also been reported in both children and adults following several other cerebellar insults, including vascular events, infections, and trauma [4].

The uncertain etiology of PFS, myriad of cited risk factors and therapeutic challenges make this phenomenon an elusive entity.

Risk Factors

Cerebellar mutism risk factors.

Posttraumatic cerebellar mutism

Cerebellar mutism is a rare occurrence following paediatric trauma [5) 6) 7) 8)]. , this phenomenon has rarely been reported following other insults, such as trauma, and its pathophysiology remains poorly understood.

A seven-year-old child who presented to the casualty department of Sultan Qaboos University Hospital in Muscat, Oman, in May 2013 with a traumatic right cerebellar contusion. The child presented with clinical features of cerebellar mutism but underwent a rapid and spontaneous recovery [9)].

Pathophysiology

Cerebellar mutism Pathophysiology.

Pathogenesis

The pathogenic mechanism is likely due to the damage occurring to the proximal efferent cerebellar pathway, including the dentate nucleus, the superior cerebellar peduncle, and its decussation in the mesencephalic tegmentum [10)].

Superior and inferior cerebellar peduncles and the superior part of the cerebellum were related to CMS, especially the right side [11)].

Clinical features

This syndrome involves a variety of signs and symptoms including cerebellar mutism or speech disturbances, dysphagia, decreased motor movement, cranial nerve palsy and, emotional lability. These signs and symptoms develop from an average range of 24 to 107 hours after surgery and may take weeks to months to resolve.

Diagnosis

Multi-inflow time arterial spin-labeling shows promise as a noninvasive tool to evaluate cerebral perfusion in the setting of pediatric obstructive hydrocephalus and demonstrates increased CBF following the resolution of cerebellar mutism syndrome [12)].

Differential diagnosis

The importance of olivary hypertrophic degeneration as a differential diagnosis in cerebellar mutism syndrome [13)].

Prevention

Cerebellar mutism prevention.

Outcome

Early recognition of this syndrome could facilitate preventive and restorative patient care, prevent subsequent complications, decrease length of hospital stays, and promote patient and family understanding of and coping with the syndrome [14].

Case series

20 cases of PFS (8%), 12 males and 8 females. Age ranged from 1.5 to 13 years (mean = 6.5). Of the 20, 16 were medulloblastoma, 3 were ependymoma and 1 was astrocytoma. There was a 21 % incidence (16/76) of PFS in medulloblastoma of the posterior fossa. The incidence for ependymoma was 13% (3/24) and 1% (1/102) for astrocytoma. All 20 cases (100%) had brainstem involvement by the tumor. The most frequent postoperative findings included mutism, ataxia, 6th and 7th nerve palsies and hemiparesis. Mutism had a latency range of 1-7 days (mean = 1.7) and a duration of 6-365 days (mean = 69.2, median = 35). Although mutism resolved in all cases, the remaining neurologic complications which characterized our findings of PFS were rarely reversible. We describe potential risk factors for developing PFS after surgery with hopes of making neurosurgeons more aware of potential problems following the removal of lesions in this area. Early recognition of PFS would further promote patient and family understanding and coping with this síndrome [15]

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19 children diagnosed with posterior fossa syndrome [16]

Case reports

Cerebellar mutism case reports.

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