

Posterior fossa syndrome

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Posterior fossa syndrome (PFS), also known as cerebellar mutism syndrome or cerebellar cognitive affective syndrome, is a rare and complex condition that can occur following surgery to remove tumors or other lesions located in the posterior fossa of the brain. The posterior fossa is the space at the back of the skull that houses the cerebellum, brainstem, and fourth ventricle.

Key features of posterior fossa syndrome include:

Onset: PFS typically occurs after surgery to treat tumors or lesions in the posterior fossa, particularly those involving the cerebellum.

Cerebellar Mutism: One of the hallmark features is the development of mutism (lack of speech) or severe reduction in speech output. Despite the term “mutism,” it's important to note that this condition may involve not only speech but also a broader range of cognitive and affective changes.

Cognitive and Affective Changes: In addition to mutism, individuals with PFS may experience a variety of cognitive and affective (emotional) changes. These can include difficulties with concentration, irritability, emotional lability, and behavioral changes.

Motor Impairments: While the cerebellum is primarily associated with motor coordination, PFS may also involve motor impairments such as ataxia (lack of coordination and balance), weakness, or other motor deficits.

Recovery: Many individuals with PFS show a degree of recovery over time, with the return of speech and improvement in other symptoms. However, the recovery can be variable, and some individuals may continue to experience long-term challenges.

The exact mechanisms underlying posterior fossa syndrome are not fully understood. It is believed that the surgical trauma, disruption of neural pathways, and possible damage to the cerebellum and surrounding structures contribute to the development of symptoms.

Management of posterior fossa syndrome involves a multidisciplinary approach, including neurology,

neurosurgery, speech therapy, and rehabilitation services. Speech therapy is particularly important in addressing the mutism and communication difficulties associated with the syndrome. Supportive care and rehabilitation efforts aim to maximize the individual's functional abilities and quality of life.

It's important to note that the prognosis and outcomes can vary widely among individuals, and the management plan is tailored to each person's specific needs and challenges. Regular follow-up and ongoing support are crucial components of care for individuals affected by posterior fossa syndrome.

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

Most posterior fossa tumors present with signs and symptoms of intracranial hypertension due to hydrocephalus (HCP).

These include:

- a) headache.
- b) nausea/vomiting: due either to intracranial hypertension due to hydrocephalus (HCP) or from direct pressure on the vagal nucleus or the area postrema (so-called "vomiting center")
- c) papilledema: estimated incidence is $\approx$ 50-90% (more common when the tumor impairs CSF circulation). Chronic increased pressure can cause irreversible blindness from the optic nerve atrophy
- d) gait disturbance/ataxia
- e) vertigo
- f) diplopia: may be due to abducens nerve palsy, which may occur with increased ICP in the absence of direct compression of the nerve

2. S/S indicative of mass effect in various locations within the posterior fossa

- a) lesions in cerebellar hemisphere may cause: ataxia of the extremities, dysmetria, intention tremor
- b) lesions of cerebellar vermis may cause: broad-based gait, truncal ataxia, titubation
- c) brainstem involvement usually results in multiple cranial nerves and long tract abnormalities, and should be suspected when nystagmus is present (especially rotatory or vertical)

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