International Journal of Advanced Science and Research
ISSN: 2455-4227
Impact Factor: RJIF 5.12
www.allsciencejournal.com
Volume 3; Issue 6; November 2018; Page No. 16-17

Bobble head doll syndrome

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Abstract
The "bobble-head doll syndrome (BHDS)" is a rare and an interesting movement disorder characterized by continuous or episodic forward and backward head nodding at times associated with side to side movements of the head of 2-3 Hz frequency. Although the exact pathogenesis of bobble-head doll syndrome is still unknown, there are many theories as to how and why it does what it does. Most of these theories acknowledge the striking similarity of symptoms between bobble-head doll syndrome and other movement disorders. The main physical symptom of bobble-head doll syndrome is the two to three bobs per second of the head, which can sometimes also include the shoulders and upper torso. Increased in stressful situations whereas they decreased in concentration (e.g. looking at a book). Endoscopic ventriculo cysto cisternostomy, is the optimal treatment option and Ventriculoperitoneal shunt is made to reduce the intracranial pressure caused by the accumulation of CSF in the third ventricle.

Keywords: bobble-head doll syndrome (BHDS), Cavum Pellucidum, Interpositum

Introduction
Bobble-head doll syndrome (BHDS) was first described by Benton *et al*. in the year 1966. It mainly affecting children under 10 years of age, is a rare and surgically treatable movement disorder characterized by bobing their head and shoulders forward and back, or sometimes side-to-side, involuntarily, in a manner reminiscent of a bobble head doll.

Definition
The "bobble-head doll syndrome (BHDS)" is a rare and an interesting movement disorder characterized by continuous or episodic forward and backward head nodding at times associated with side to side movements of the head of 2-3 Hz frequency

Causes
- Lesion In and Around the Third Ventricle.
- Third Ventricular Tumors or Suprasellar Arachnoid Cysts.
- Aqueductal Stenosis.
- Hydrocephalus
- Cyst Of the Cavum Pellucidum And Interpositum,
- Trapped Fourth Ventricle,
- Aqueductal and Third Ventricular Choroid Plexus Papilloma.
- Developmental Cerebellar Disorders.

Pathophysiology
Although the exact pathogenesis of bobble-head doll syndrome is still unknown, there are many theories as to how and why it does what it does. Most of these theories acknowledge the striking similarity of symptoms between bobble-head doll syndrome and other movement disorders.
The presence of cystic lesions, causing swelling in the third ventricle, is a common feature in all patients. It is this dilatation that causes pressure to be applied to the surrounding structures of the third ventricle, such as the diencephalon. It is possible that the back and forth movement of fluid within the cyst causes rhythmic pressure on the diencephalic motor pathways.
One of the key periventricular structures in that pathway is the thalamus which is responsible for relaying motor signals to the cerebral cortex as well as regulating consciousness, sleep, and alertness. The tracts associated with the extrapyramidal system are controlled by various structures of the central nervous system, such as the cerebellum and basal ganglia. The basal ganglia plays a large part in controlling motor function and thus, abnormalities to this system can result in movement disorders such as Parkinson's Disease and dyskinesia, both of which share commonalities with bobble-head doll syndrome

Signs and symptoms
The manifestations of bobble head doll syndrome is categorized into three.

Physical
- The main physical symptom of bobble-head doll syndrome is the two to three bobs per second of the head, which can sometimes also include the shoulders and upper torso. Increased in stressful situations whereas they decreased in concentration (e.g. looking at a book). It was more and more a strain that the no-no direction of the movement was interpreted as a negative communication by others causing misunderstandings.
- The patient is unaware of the movements and unable to control them unless directed to stop or given simple mental tasks such as basic arithmetic or spelling words.
- A supplemental symptom of the head bobbing is a presence of ataxia, difficulty walking, running, and climbing steps because of the bobbing.

- Obesity.

Neurological
- An enlargement of the head due to accumulation of cerebrospinal fluid in the third ventricle.
- Rhythmic head extension
- Rhythmic head flexion
- Rhythmic arm extension
- Rhythmic arm flexion
- Fine tremor
- Mental retardation
- Impaired vision
- Hydrocephalus

Psychiatric symptoms
- Elated mood
- Talkativeness
- Inflated self-esteem
- Distractibility
- Intermittent aggressiveness

Diagnostic Evaluations
- Cisternography to observe obstruction in cerebrospinal fluid (CSF) flow among ventricles.
- Mid-sagittal plane MRI is the best modality for the delineation of CSF pathways and soft tissue.
- A CT Scan is an alternative to isternography which is used to see CSF flow and might be useful in planning choice of surgery.

Management

Successful surgical procedures include
- Surgical Removal Of The Lesion
- Ventriculoperitoneal Shunt
- Endoscopic Ventriculocisternostomy
- Transcallosal Cystectomy

Removal of lesion

In the case of choroid plexus papilloma, surgical removal of the cyst-containing lesion from within the third ventricle caused a full recovery, the mobile nature of the cystic lesion led to its intermittent obstruction of the foramen of Monro and proximal aqueduct, producing the bobble-head symptoms. Once removed, all symptoms disappeared.

Ventriculoperitoneal shunt

Ventriculoperitoneal shunt is made to reduce the intracranial pressure caused by the accumulation of CSF in the third ventricle. Typically, this will succeed in restricting the swelling and allowing proper flow of CSF, with this relief, the head bobbing will disappear and bobble-head doll syndrome will no longer be present.

Endoscopic ventriculo cysto cisternostomy

Child with suprasellar arachnoid cysts, undergoing endoscopic ventriculo cysto cisternostomy, is the optimal treatment option. By fenestrating, or opening, the cystic membrane and removing the fluid, all obstructions of the aqueduct were resolved. In patients receiving this treatment, a full recovery is the most common result.

Transcallosal Cystectomy

A cyst within the 3rd ventricle was excised sub totally by a transcallosal approach. Postoperatively it reduces head bobbing and improves memory.

Conclusion

The bobble-head doll syndrome (BHDS) is characterized by a back-and-forth movement of the head with a frequency of 2 to 3 Hz, which increases during walking and excitement and decreases during concentration. The head movements are accompanied by macrocephaly, ocular disturbances, psychomotor retardation, and sometimes endocrine dysfunction. Cisternography and MRI are the main diagnostic methods for diagnosing this syndrome. endoscopic ventriculo cysto cisternostomy and Transcallosal Cystectomy are the major surgical procedures carried out to reduce head bobbing and improves memory in affected children.

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