JOURNAL OF AMERICAN MEDICAL SCIENCE & RESEARCH

Mini Review
ISSN: 3041-5578
Journal homepage: https://medical.researchfloor.org/

Bobblehead Syndrome- A Systematic Review

***Essra Ali Safdar and Nida Ali Safdar**

Department of Pharmacy Practice, Anwarul Uloom College Of Pharmacy, New Mallepally Hyderabad, Telangana 500001, India

ABSTRACT

Bobblehead Syndrome (BHS) is a rare neurological condition characterized by rhythmic and involuntary head movements, resembling a bobbing motion. This systematic review aims to consolidate current knowledge on the etiology, clinical presentation, diagnostic methods, and treatment options for BHS. A comprehensive search of peer-reviewed articles from medical databases was conducted, focusing on case reports, clinical studies, and reviews published over the past three decades. Findings indicate that BHS is frequently associated with third ventricular cysts, particularly colloid cysts, or hydrocephalus. Diagnostic imaging, such as magnetic resonance imaging (MRI) and computed tomography (CT), plays a crucial role in identifying underlying anatomical abnormalities. Treatment often involves surgical intervention, including ventriculoperitoneal shunting or endoscopic cyst fenestration, which significantly reduces symptoms in most cases. Despite advancements in diagnostic and therapeutic modalities, the rarity of BHS poses challenges in early recognition and management. This review underscores the need for increased awareness and further research to optimize outcomes for affected individuals.

Keywords: *Bobblehead Syndrome, third ventricular cysts, hydrocephalus, rhythmic head movements, neurological disorders*

Citation: Essra Ali Safdar and Nida Ali Safdar (2024). Bobblehead Syndrome- A Systematic Review. *Journal of American Medical Science and Research.*

DOI: https://doi.org/10.51470/AMSR.2024.03.02.23

Received on: 12 August, 2024
Revised on: 09 September, 2024
Accepted on: 25 October, 2024

Corresponding Author: ***Essra Ali Safdar***
Email Address: ***aliisra638@gmail.com***

Copyright: *© The Author(s) 2024. This article is Open Access under a Creative Commons Attribution 4.0 International License, allowing use, sharing, adaptation, and distribution with appropriate credit. License details: http://creativecommons.org/licenses/by/4.0/.*
Data is under the CC0 Public Domain Dedication (http://creativecommons.org/publicdomain/zero/1.0/) unless otherwise stated.

Introduction

Benton and associates first described Bobble Head Syndrome in 1966. Bobble Head Syndrome (BHS)is a rare pediatric neurological condition typified by involuntary, repetitive head movements that are rarely horizontal (no - no) and frequently up and down (yes - yes) with a frequency of 2-3 hz. They are rhythmic and stereotypical, and they get worse while under stress or emotion. They get better when tilting their head back, moving involuntarily, or sleepin [1]. Due to the condition's rarity and maybe underdiagnosis of the illness itself, epidemiology is unknown. It primarily affects children under the age of three.

BHS is usually associated with a third ventricle tumor, aqueductal stenosis, suprasellar arachnoid cysts, a third ventricle cystic choroid plexus papilloma, a trapped fourth ventricle, a cavum septum pellucidum cyst, and other lesions around the third ventricle [2].

The syndrome includes the movement condition as well as other symptoms such as macrocephaly, ataxia, developmental delay, optic disk pallor or atrophy, hyperreflexia, tremor, obesity, endocrine issues, headache, and vomiting. Neuroimaging is essential for both diagnosis and postoperative monitoring. In order to treat bobblehead doll syndrome, which is caused by a cyst in the third ventricle, various surgical techniques have been described. These include shunting the cyst, a subfrontal approach with ablation of the anterior wall of the cystic lesion, and, more recently, the minimally invasive procedure by neuro-endoscopy, which involves fenestration of the cyst wall [3-1].

Pathophysiology

The primary symptom of the complicated illness known as bobblehead doll syndrome is repetitive anteroposterior head movement. The dilated third ventricle's pressure impact is thought to be the cause of this movement condition because it causes distortions in the dorsomedial red nucleus and dentatorubrothalamic pathway [4-6]. Furthermore, the medial thalamus, which has a somatotropic motor representation of the head and neck region, is compressed [6]. Tremors of the head and neck are caused by pressure over the medial thalamus, and when the lesion worsens, truncal and appendicular tremors are also caused by the thalamic area being affected. Tremors' existence at rest and attenuation during volitional activity point to a potential basal ganglia role in the development of this illness. Additionally, as the movement halts on volitional acts, it is hypothesized that it develops as a learned behavior. The fact that head motions and headaches stop when the neck is flexed or extended to an excessive degree in a BHDS case suggests that this aids in increasing the circulation of CSF [7].

Clinical presentation

The primary clinical characteristic is the head's bobbling, which is described as nodding occasionally connected to side-to-side head motions of 2-3 Hz frequency. When given or forced to perform duties, these movements can occasionally be made to stop, and this is best understood in the early phases. Since it is more consistent in nature, this is not acknowledged in the circumstances of people with major pathologies or late presentations. Ataxia may accompany the bobbling. Patients who have obstructed hydrocephalus frequently exhibit big heads and other hydrocephalus symptoms because of blockage at the level of the foreman of Monro [8]. Static and dynamic ataxia can severely impede a patient's ability to walk straight or climb stairs, and dysdiadochokinesia is occasionally observed. Some patients experience insomnia. Bulimia and other eating disorders are sometimes mentioned during the course of the illness. It appears that diseases of the optic nerve and/or visual pathway are the cause of the reported decreased visual acuity. Additional symptoms of the illness include significant intellectual impairment, developmental delay, and attention deficit. In addition to the potential diagnosis of enuresis, developmental abnormalities such as delayed puberty, small stature (below the third percentile), or early puberty with markedly high testosterone levels may also manifest [9].

©2024 Journal of American Medical Science and Research. All Rights Reserved.

Diagnosis

Brain magnetic resonance imaging (MRI) is the primary diagnostic technique for the condition mentioned. Different pathological alterations in the brain are described, despite a comparable clinical presentation. The picture can reveal obstructive hydrocephalus and an arachnoid cyst in the third ventricle's suprasellar area. The condition may occasionally be caused by a thin-walled cyst in the sellar and suprasellar region. The fourth ventricle may subsequently grow as a result of an increasing cyst during the course of Bobble-Head Doll Syndrome. A cyst of the septum pellucidum may occasionally be found. Less commonly, pedunculated cystic alterations in the anterior region of the third ventricle close to the Monro foramen may be seen on MRI along with concurrent enlargement of the lateral and third ventricle.(10) When evaluating CSF flow, MR imaging is also crucial. CSF flow towards the cyst can occasionally be seen in the MR FLAIR sequence. Although brain MRI is currently conducted considerably more frequently due to its less invasive nature and improved accuracy in detecting alterations, computed tomography (CT) of the head is another important diagnostic technique. One important test that seems to be useful for assessing CSF flow is CT cisternography. A further diagnostic technique is electromyography (EMG). The EMG analysis of muscles like the trapezius allows for the detection of head bobbing [11,9].

Treatment

The primary treatment for the underlying cause of Bobble-Head Doll Syndrome is neurosurgical intervention. Usually, symptoms go away on their own once the cyst is removed. In the past, the main therapeutic approaches were open marsupialization and permanent ventriculoperitoneal or cystoperitoneal drainage [12]. Most patients seem to respond well to endoscopic fenestration of the cyst with Ventriculocystocisternostomy, but in certain instances, it might be deemed impossible because of the potential for adverse anatomical conditions or the risk of damage to the basal cerebral artery [13]. Sometimes the placement of a cystoventriculoperitoneal shunt is performed under neuroendoscopic monitoring, especially if the cyst is closely associated with hydrocephalus, which raises intracranial pressure. Nevertheless, using this valve comes with a higher risk of issues like infection.

Conclusion

Bobblehead syndrome is a complex neurological disorder with unknown etiology. The time between diagnosis and surgery appears to be a predictor of the clinical result. The pathogenesis of the illness is probably complex and is still not fully understood. Because of its positive results and low risk of problems, endoscopic ventriculo-cisterno-stomy is the recommended course of treatment.

Acknowledgements

The authors express their gratitude to the patient who participated in this study for their cooperation and trust.

Funding: This research received no external funding.

Availability of Data and Materials

The data and materials used in this study are available from the corresponding author upon reasonable request.

Ethics Approval and Consent to Participate

This study was conducted in accordance with the Declaration of AUC, Hyderabad, and received approval from the Department of Pharmacy Practice, Anwarul Uloom College of Pharmacy, New Mallepally, Hyderabad, Telangana, India (500001). The patient was fully informed about the study, and informed consent was obtained in their native language before publication decisions were made. Documentation of consent has been provided at the time of submission.

Consent for Publication

Written informed consent was obtained from the patient for the publication of this case report and any accompanying images. A copy of the signed consent is available for review with Journal.

Competing Interests

The authors declare that they have no inancial or non-inancial competing interests.

References

1. Mohamed Dahamou Et Al Bobble head doll syndrome (BHDS): Case report Radiology Case Reports 18 (2023) 2498–2502.
2. Bhattacharyya et Al Bobbling head in a young subject DOI: 10.4103/0972-2327.143992.
3. Mohamed Dahamou Et Al Bobble head doll syndrome (BHDS): Case report Radiology Case Reports 18 (2023) 2498–2502.
4. Coker SB. Bobble-head doll syndrome due to trapped fourth ventricle and aqueduct. Pediatr Neurol. 1986;2:115–6. doi: 10.1016/0887-8994(86)90069-x. [DOI] [PubMed] [Google Scholar]
5. Dell S. Further observations on the “Bobble-Headed doll syndrome”. J NeurolNeurosurg Psychiatry. 1981;44:1046–9. doi: 10.1136/jnnp.44.11.1046. [DOI] [PMC free article] [PubMed] [Google Scholar]
6. Russo RH, Kindt GW. A neuroanatomical basis for the bobble-head doll syndrome. J Neurosurg. 1974;41:720–3. doi: 10.3171/jns.1974.41.6.0720. [DOI] [PubMed] [Google Scholar]
7. Wiese JA, Gentry LR, Menezes AH. Bobble-head doll syndrome: Review of the pathophysiology and CSF dynamics. Pediatr Neurol. 1985;1:361–6. doi: 10.1016/0887-8994(85)90073-6. [DOI] [PubMed] [Google Scholar]
8. N Zimani Et al A Rare Case of Bobble Head Doll Syndrome Current Opinions in Neurological Science ISSN: 2575-5447.
9. Piotr Gacka Et Al Bobble-Head Doll Syndrome: Understanding a Rare Neurological Enigma DOI:10.12775/JEHS.2024.69.55355.
10. Ishihara M Et Al "No-no" type bobble-head doll syndrome in an infant with an arachnoid cyst of the posterior fossa: a case report. Pediatr Neurol. 2013;49(6):474 -476. doi:10.1016/j.pediatrneurol.2013.07.013.
11. Piotr Gacka Et Al Bobble-Head Doll Syndrome: Understanding a Rare Neurological Enigma DOI:10.12775/JEHS.2024.69.55355.
12. Guerreiro H et al Bobble-head doll syndrome: therapeutic outcome and long-term follow-up in four children. Acta Neurochir(Wien).2012;154(11):2043-2049. doi:10.1007/s00701-012-1458-2.
13. Hagebeuk EE Et Al Bobble-head doll syndrome successfully treated with an endoscopic ventriculocystocisternostomy. Case report and review of the literature. J Neurosurg. 2005;103(3 suppl);253-259. doi:10.3171/ped.2005.103.3.0253.

©2024 Journal of American Medical Science and Research. All Rights Reserved.