Bobble-head doll syndrome

Bobble-head doll syndrome is a rare movement disorder that is usually associated with lesions involving the third ventricle. It is characterised by stereotypical rhythmic up-and-down or side-to-side head movements. The pathophysiology and anatomical basis for this unusual manifestation is still a subject of intense scrutiny.

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a 1.5-year-old Asian-Syrian girl who presented with a history of excessive head nodding for 3 months that increased with walking, emotions, and stress; decreased during periods of concentration; and was absent during sleep. On physical examination, she was alert and normal, with no medical history. Laboratory assessment and ophthalmological examination were normal. Cranial magnetic resonance imaging demonstrated a well-defined thin-walled suprasellar arachnoid cyst measuring 3 × 5 × 7 cm that obstructed the foramina of Monro, with resulting hydrocephalus ventriculomegaly. The patient underwent endoscopic cystoventriculostomy and cystocisternostomy for the suprasellar arachnoid cyst. During the 6 months of follow-up, the head bobbing disappeared completely, and her growth was normal.

Despite the rareness of bobble-head doll syndrome, it is considered an important condition that must be investigated early to detect the cause and treated promptly to avoid potential complications [1)].

The syndrome has never been described in a patient with both hydranencephaly and Chiari type 3 malformation.

We describe a 2-year-old female patient who presented with congenital hydrocephalus, an occipital encephalocele and rhythmic bobbling of the head. Imaging investigation revealed a Chiari type 3 malformation and hydranencephaly. The patient was taken to theatre for a ventriculoperitoneal shunt insertion, and at day 3 post operatively, the patient had a markedly decreased head circumference and a decrease in the frequency of the bobbling of the head. A further review at 2 weeks showed that the bobbling of the head had ceased. Although the pathophysiology of bobble-head doll syndrome is yet to be fully understood, there has been postulation of either a third ventricular enlargement or a cerebellar dysfunction to explain bobble-head doll syndrome. Our case illustrates that the pathophysiology is most likely multifactorial as illustrated by the fact that by just addressing the high intracranial pressure with a shunt was sufficient to treat the condition [2)].

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Doya LJ, Kadri H, Jouni O. Bobble-head doll syndrome in an infant with an arachnoid cyst: a case report. J Med Case Rep. 2022 Oct 28;16(1):393. doi: 10.1186/s13256-022-03623-0. PMID: 36303240; PMCID: PMC9615396.
2)
Mba SE, Musara A, Kalangu K, Nyamapfene B. An unusual presentation of bobble-head doll syndrome in a patient with hydranencephaly and Chiari 3 malformation. Childs Nerv Syst. 2019 Feb 6. doi: 10.1007/s00381-019-04054-x. [Epub ahead of print] PubMed PMID: 30726525.