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CASE REPORT

# An unusual presentation of bobble-head doll syndrome in a patient with hydranencephaly and Chiari 3 malformation

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## Abstract

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. 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.

**Keywords** Bobble-head doll · Hydranencephaly · Chiari type 3

## Introduction

Chiari type 3 is the rarest form of Chiari malformations [28]. It is described as herniation of hindbrain elements into an encephalocele. Chiari type 3 has been associated with various other developmental abnormalities but has not been described to occur in a patient with hydranencephaly.

Bobble-head doll syndrome is described as a rare neurological stereotypic head movement disorder characterised by about 2 to 3 Hz of periodic anterior-posterior and occasionally side-to-side head movements [4, 11, 17, 20, 22]. It was first described by Benton et al. [3] in 1966 and has been invariably associated with third ventricular expansion [9, 19, 29], either by a cyst or a stenosed aqueduct of Sylvius, etc.

This is the first reported case of a patient with a combination of rare pathologies; bobble-head doll, hydranencephaly and Chiari 3 malformation.

## Case report

A 2-year-old female patient (Figs. 1, 2) presents to the neurosurgeon with abnormal movements of the head for the past year. She had been lost to follow-up when she was diagnosed with congenital hydrocephalus and occipital encephalocele at birth.

The head movements were repetitive and rhythmic, about 75 movements/min, and were said to have started at 1 year of age.

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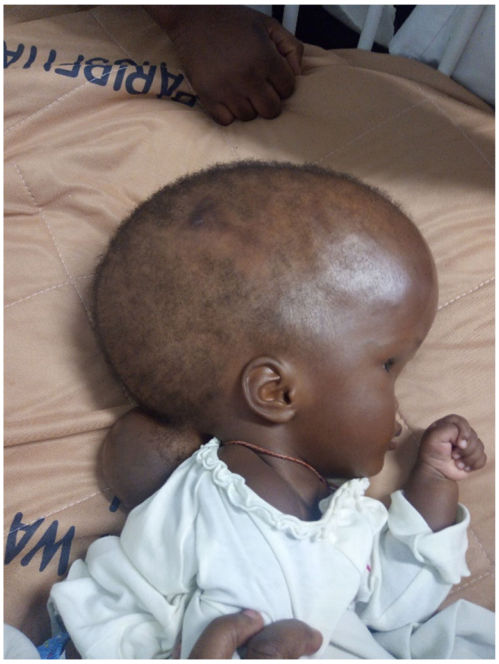

**Fig. 1** Child with macrocephaly and occipital encephalocele

The mother's pregnancy was noted to be uneventful, with no known perinatal infection.

Patient had significant developmental milestone delay but no history of seizure or vomiting.

Her immunisation record was up to date and the patient tested HIV (human immunodeficiency virus) negative.

On examination, she was blind and small for her age with an occipito-frontal circumference of 73 cm (above 2 standard deviations from normal) and had dysmorphic features (hypertelorism, low-set ears), distended scalp veins and parinaud syndrome.

She had no neck stiffness, and an occipital encephalocele measuring 6 × 4 × 5 cm was noted. No other abnormalities were noticed in the cardiovascular or gastrointestinal system.

**Fig. 2** A still image from the video showing bobble-head doll syndrome prior to ventriculoperitoneal shunt insertion (video can be found in the supplementary data of this article)

Imaging investigations showed hydranencephaly and Chiari 3 malformation (Fig. 3).

A ventriculoperitoneal shunt was inserted and the repair of the occipital encephalocele was scheduled for a later date.

The patient responded well to the shunt procedure at 72 h with an initial decrease in the size of the head (from 73 to 62 cm) and a decrease in the frequency of head bobbling (from 75 to 40 movements/min).

At 2-week follow-up, the head bobbling had ceased.

## Discussion

This case presentation is peculiar at various levels, not only because the collection of symptoms is rare but also because of the positive response observed after ventriculoperitoneal shunt insertion.

There is no unanimity about the pathophysiological process underlying bobble-head doll syndrome [22]. Russo et al. [23] gave a neuroanatomical basis that involves pressure from the third ventricle transmitted to the dorsomedial nucleus of the thalamus [22], from there, the pathway course to the lentiform nucleus and the prerubral fields of Forel; this in turns connects to the midbrain tegmentum and red nucleus and project onto the motor neurones in the cervical spinal cord (Fig. 4).

The above explanation however is not sufficient as patients without ventricular dilatation also present with bobble-head doll syndrome; in those patients, there is postulated to be an association between posterior fossa malformations and bobble-head doll syndrome [5, 12, 25]. This association is also evident in our patient with Chiari 3 malformation.

Some authors still believe the repetitive movements of the head are a learned response meant to help improve cerebrospinal fluid circulation [21, 27].

The many reported cases of bobble-head doll syndrome in the literature are an eclectic collection of associated features with arachnoid cyst dominating as a feature [10] but no typical causative factor identified [1, 2, 7, 8, 13, 18, 24, 26].

The triple association of Chiari 3 malformation, hydranencephaly and bobble-head doll syndrome has not been described in the literature. These probably represent various stages of developmental abnormalities; hydranencephaly is a post-neurulation defect that is the result of either congenital infection or bilateral internal carotid infarcts [6, 15, 16]. It is possible that a Chiari 3 will develop following changes in CSF dynamics [14] as purported in our patient; it is far however to acknowledge that the origin of this collection of rare occurrences is still unknown.

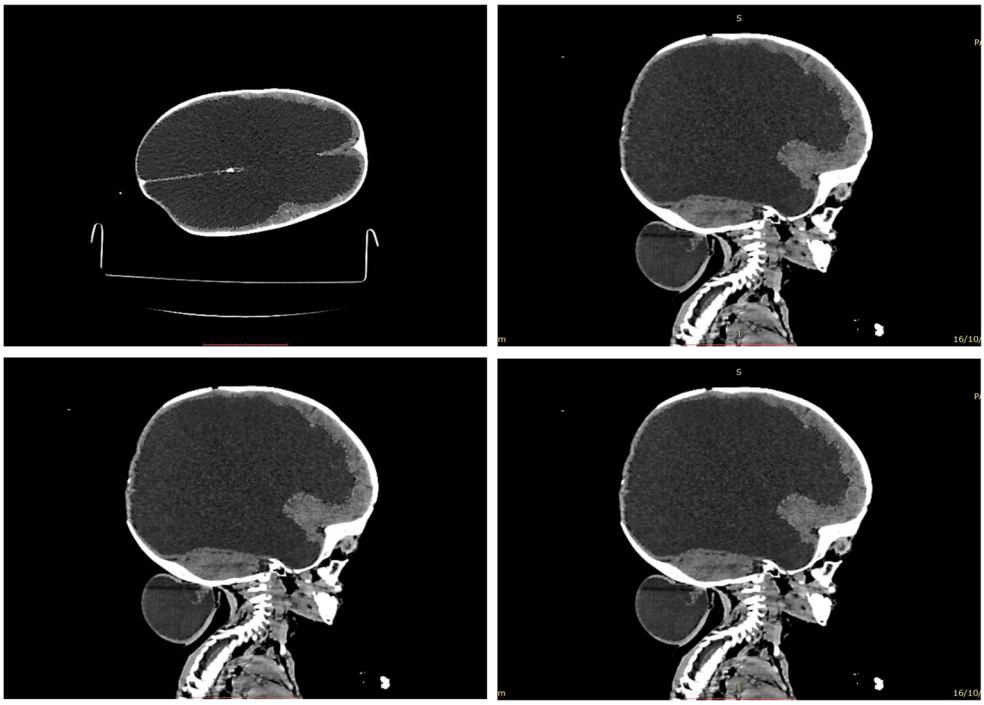

**Fig. 3** Sagittal CT scan showing hydranencephaly and occipital encephalocele representing a Chiari 3 malformation

## Conclusion

The occurrence of bobble-head doll syndrome in a patient with hydranencephaly and Chiari malformation type 3 has not been reported before. The resolution of symptoms after ventriculoperitoneal shunt insertion may point towards a different aetiological factor in the development of bobble-head doll syndrome. Whatever the pathogenesis basis, one needs to consider raised intracranial pressure amongst the aetiological factors.

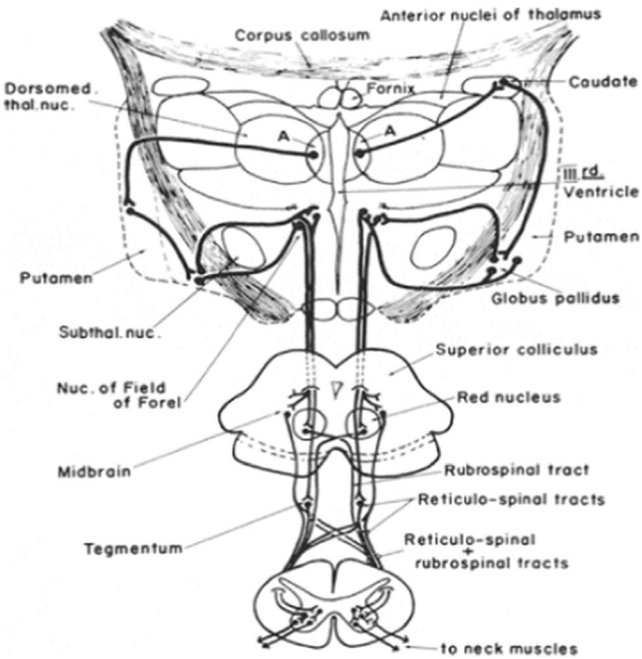


**Fig. 4** Discharge pathway from the dorsomedial thalamic nucleus (adapted from [23])

**Compliance with ethical standards** The authors declare that the study complies with the current law in Zimbabwe.

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**Conflict of interest** The authors declare that they have no conflict of interest.

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