A neuroanatomical basis for the bobble-head doll syndrome

R. Harris Russo, M.D., and Glenn W. Kindt, M.D.

Department of Surgery, Section of Neurosurgery, University of Michigan Medical Center, Ann Arbor, Michigan

A case of the bobble-head doll syndrome in association with aqueductal stenosis is presented. The relationship between the dilated third ventricle and adjacent thalamic nuclei as well as the somatotopic motor pattern of the dorsomedial thalamic nucleus is discussed.

Key Words • hydrocephalus • bobble-head doll syndrome • basal ganglion • dorsomedial thalamic nucleus • third ventricle • aqueductal stenosis

The "bobble head doll syndrome" exhibits a two-to-three per second flexion-extension of the head and neck on the trunk associated with third ventricular dilatation from a cyst or hydrocephalus. We are reporting the second case in the literature associated with aqueduct stenosis. We propose a neuroanatomical explanation for the syndrome.

Case Report

A 1-year-old girl was admitted to the Mott Children's Hospital because of abnormal movements of an enlarging head. She was the first child of a healthy 19-year-old mother, and the product of a normal pregnancy and delivery. She weighed 3300 gm at birth and had a normal head size. At 2 months, she was treated for H. influenza meningitis, and developed an enlarging head over the ensuing 3 months. Bobbing movements of the head began at the age of 5 months.

Examination. The child was lethargic and had no interest in her surroundings. She exhibited psychomotor retardation and did not speak or crawl. The head measured 51.5 cm with prominent frontal bossing and bulging anterior and posterior fontanelles. Her height was 73 cm and weight 9.76 kg. Blood pressure was 125/80 and pulse 120. Neurological examination revealed increased muscle tone and hyperactive reflexes in both legs; the arms were normal. The head was constantly moving backward and forward. The arms were not involved although the movements flowed in an undulating fashion into the shoulders. The bobble ceased when the child reached for an object, and during sleep, but increased in amplitude during excitement. The child could stop the motion on command. Laboratory studies were normal. Skull films showed a cranial capacity above the 95th percentile. An electromyogram showed contractions of the cervical erector spinae muscles at the rate of two-to-three per second (Fig. 1). Air ventriculography revealed dilated lateral and third ventricles due to aqueductal obstruc-
Bobble-head doll syndrome

The third ventricle appeared ballooned more than is usually seen with aqueductal stenosis (Fig. 2). Testing of the cerebrospinal fluid revealed that glucose was 47 mg% and the total protein 18 mg%; there were 55 red blood cells and 4 mononuclear cells/cu mm. 

Operation. A right ventriculoatrial Holter shunt was placed. At operation the intraventricular CSF pressure was 350 mm H₂O. 

Postoperative Course. Postoperatively the child was brighter and more spontaneous. The head bobble disappeared on the eighth postoperative day. She was crawling and beginning to speak at the sixth month follow-up examination. The head circumference was stable at 50 cm, and there has been no recurrence of the head bobble.

Discussion

To-and-fro movements of the head were described by Wilson² as a symptomatic rhythmia seen in a variety of neurological disorders. He defined rhythmia as a “coordinated and seemingly purposeful movement repeated monotonously and often attended with pleasurable feeling-tone.” Benton, et al.,¹ first used the term “bobble-head doll syndrome” when describing children with third ventricular cysts who exhibited the to-and-fro head movements; several case reports have appeared since.⁵⁷

The common denominator in all cases reported is third ventricular dilatation. Pneumoencephalographic studies in adults with a variety of movement disorders disclosed statistically significant enlargement of the third ventricle in comparison to a control group.⁶ Endocrine abnormalities have been reported in hydrocephalus,⁸ presumably a result of third ventricular enlargement with compression of hypothalamic nuclei. Whether in our case the dilated third ventricle caused irritation or depression of subcortical neuronal arcs is not known. The pathophysiologic mechanism appears reversible as the head bobble ceases with ventricular shunting procedures.

The dorsomedial nucleus of the thalamus...
occupies a large paraventricular area that would be compressed with dilatation of the third ventricle. Stimulation studies have shown a somatotopic motor pattern in this nucleus, with the legs most rostral and lateral, and the head and neck areas most caudal and medial. This pattern has recently been confirmed in our laboratory. The discharge pathways from the dorsomedial thalamic nucleus to the motoneurons of the cervical spinal cord can be described by known projections.

The dorsomedial nucleus projects laterally to the cerebral hemisphere and the subfrontal regions, as well as to the caudate and the putamen. Pathways course from the caudate and putamen to the globus pallidus, and through the lenticular fasciculus and ansa lenticularis to the prerubral fields of Forel. Polysynaptic pathways from the fields of Forel enter the midbrain tegmentum and red nucleus from which crossed and uncrossed rubroreticulospinal and reticulospinal tracts descend to motoneurons in the cervical spinal cord (Fig. 3).

The anterior nucleus of the thalamus is also adjacent to the third ventricle but probably is not involved in the genesis of head bobble. Fibers from the medial mammillary nucleus of the posterior hypothalamus constitute the main input to the anterior nuclear group. This nucleus projects through the anterior thalamic radiations to the cingulate gyrus. Stimulation of the cingulum causes autonomic and motor excitation, but stimulation of the anterior thalamic nucleus in our laboratory revealed no motor pattern but resulted in arousal.

The disappearance of head bobble during volitional movement and sleep is similar to that observed in basal ganglion dyskinesias. This similarity suggests that the motor cortex is interrelated with the abnormal subcortical impulses. We suggest that these abnormal impulses originate from the medial aspect of the dorsomedial thalamic nucleus, relay to the basal ganglia and from there to motor centers.

References
Bobble-head doll syndrome


Address reprint requests to: R. Harris Russo, M.D., Department of Surgery, Section of Neurosurgery, University of Michigan Medical Center, Ann Arbor, Michigan 48104.