BOBBLE-HEAD DOLL AND DANDY-WALKER SYNDROMES

A female infant with macrocephaly (head circumference >95th%), hydrocephalus, and Dandy-Walker syndrome, who developed horizontal head movements of the ‘no-no’ type at 1 year of age, is reported from Federal University of Minas Gerais, and other centers in Brazil. CT scan on day 1 of life showed a posterior fossa cyst, hypoplasia of the cerebellar vermis, elevation of the occipital lobe, and an increase in size of the posterior fossa, findings compatible with the Dandy-Walker syndrome. With progressive enlargement of the head, a VP shunt was inserted on day 9. She developed normally in the first year and walked at 14 months. The head circumference was at the 5th percentile, and an MRI confirmed the cerebellar vermis hypoplasia and cyst, without signs of compression, enlargement or displacement of third or fourth ventricles or brainstem. Withdrawal of fluid from the fourth ventricle resulted in no change of the doll’s head movements. At 3 year follow-up, head movements persisted, but the neurological examination and milestones of development were otherwise normal. Movements were worse when walking or with stress and they disappeared in sleep. (Henriques JG de B, Henriques KSW, Filho GP, Fonseca LF, Cardoso F, Da Silva MC. Bobble-head doll syndrome associated with Dandy-Walker syndrome. J Neurosurg (3 Suppl Pediatrics) Sept 2007;107:248-250). (Reprints: Jose Gilberto de Brito Henriques MD, email: henriques-jgb@hotmail.com)

COMMENT. The case-report suggests that bobble-head doll syndrome (BHDS) may occur with cerebellar malformation, and hydrocephalus with third ventricle dilatation is not an essential causative factor. Movements are usually up and down (‘yes-yes’ type), and the ‘no-no (side-to-side) movements seen in this case are rarely reported. The most common cause is a tumor or cyst and dilatation of the third ventricle, and movements often cease after ventricular drainage. Other causes include suprasellar arachnoid cyst, and aqueductal stenosis with hydrocephalus. Age of onset varies from 2 months to 13 years. The mechanisms suggested for BHDS include pressure on the medial aspect of the dorsomedial thalamic nucleus, intermittent obstruction of the formaen of Monro, or compression of the red nucleus. Malformation of the cerebellum, especially involving the vermis, may also be involved.

INFANT DEVELOPMENTAL MILESTONES AND COGNITIVE OUTCOME

The relationship between infant developmental milestones and later intellectual function was determined in a representative sample of 5,362 children born in the United Kingdom in 1946, after the second world war, and followed in to adulthood by researchers at the Behavioral and Clinical Neuroscience Institute, University of Cambridge; and University College London, UK. When participants reached 2 years of age, mothers were asked about ages of standing and walking, and saying words. Information on at least one cognitive test (IQ at age 8, reading comprehension at age 26, or verbal fluency at age 53) and developmental variables was available in 3,969 subjects. Linear modeling demonstrated that earlier motor and speech development were significantly associated with greater IQ at age 8, higher reading comprehension at age 26, and better verbal fluency at age 57. For every month earlier in learning to stand, the individual gained one half of one IQ point at age 8. Earlier development in speech and motor milestones was also associated with better reading