

Navigated laser-assisted endoscopic fenestration of a suprasellar arachnoid cyst in a 2-year-old child with bobble-head doll syndrome

Case report

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✓The authors present the case of a 2-year-old boy with bobble-head doll syndrome (BHDS) associated with a large suprasellar arachnoid cyst and enlarged ventricles, who was successfully treated with neuronavigated laser-assisted endoscopic ventriculocystocisternostomy.

The clinical history, surgical treatment, and clinical follow up of the patient are described. A navigated laser-assisted endoscopic ventriculocystocisternostomy of the suprasellar arachnoid cyst led to cessation of the head bobbing, and notable reduction of the cyst and ventricles was visible on the postoperative magnetic resonance images.

Caused by a suprasellar arachnoid cyst, BHDS can be successfully treated with navigated laser-assisted endoscopic ventriculocystocisternostomy. The advantages of this procedure are minimal invasiveness and facilitated guidance of the neuronavigation system to the target area when normal anatomical landmarks are not visible.

KEY WORDS • bobble-head doll syndrome • suprasellar arachnoid cyst • hydrocephalus • endoscopic treatment • neuronavigation • laser treatment • pediatric neurosurgery

BOBBLE-HEAD doll syndrome is a rare movement disorder first described by Benton, et al.,² in 1966. At least 50 additional cases have been reported in the literature. The syndrome is characterized by 2- to 3-Hz rhythmic forward and backward head nodding, similar to that seen in dolls whose weighted heads rest on a coiled spring. The head bobbing is aggravated by stressful situations, diminished during voluntary movements and recumbence, and absent during sleep. The syndrome is most commonly described in children younger than 5 years of age, but it has also been reported in older children and in one adult.^{2,3,10,12,14,17,19,20,22,23,34,36}

In most cases, BHDS has been found to be associated with a cyst in the region of the third ventricle,^{1,2,9,10,12,14,20,23,34,36} and less often with aqueductal stenosis, a cyst in the cavum velum interpositum or in the cavum septum pellucidum, a cystic choroid plexus papilloma of the third ventricle, or a trapped fourth ventricle.^{3,7,12,25}

Reported treatment options for BHDS caused by a cyst in the region of the third ventricle include shunt insertion,

Abbreviations used in this paper: BHDS = bobble-head doll syndrome; MR = magnetic resonance.

endoscopic fenestration, and cyst fenestration or resection after craniotomy.^{1–3,9,10,12,14,20,21,23,24,34,36}

We report on a 2-year-old boy with BHDS who was successfully treated by a navigated laser-assisted endoscopic fenestration of a suprasellar arachnoid cyst.

Case Report

Presentation and Examination. This 2-year-old boy was referred to our hospital because of delayed motor development and abnormal head movements. The movements had started at the age of 12 months and had gradually increased. They were more pronounced in stressful situations, decreased during voluntary movements, and absent during sleep. A neurological examination revealed continuous 2-Hz to-and-fro bobbing of the head, which increased when the boy became excited. His mental and language development were normal. His head circumference at the age of 2 years was 52 cm (just below the 97th percentile). An MR image showed a large suprasellar cyst in the third ventricle with compression of the fornix, corpus callosum, pons, and mesencephalon, along with enlarged lateral ventricles (Fig.

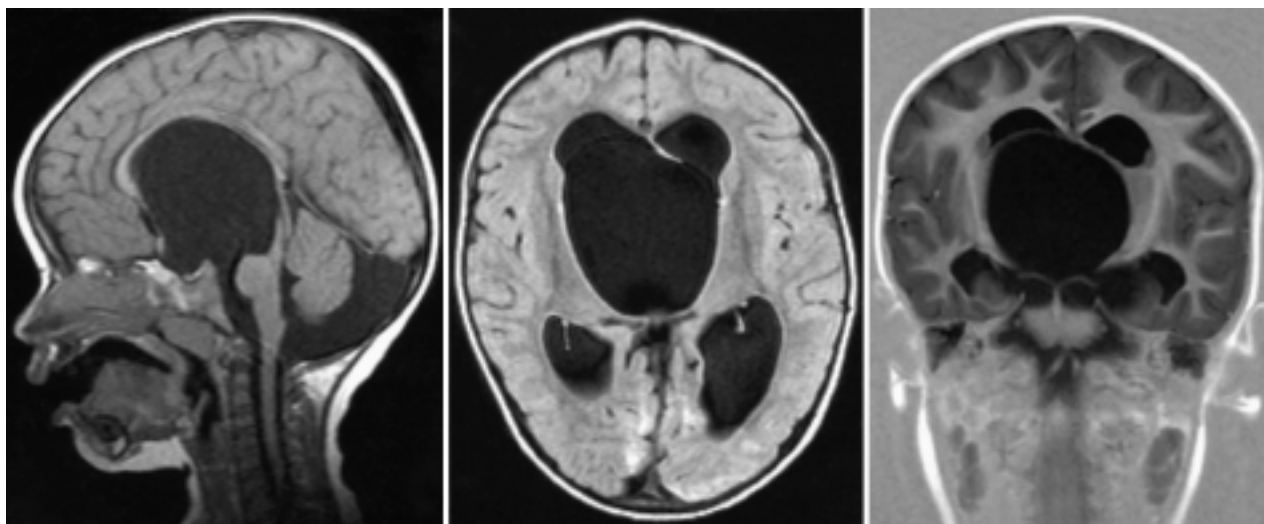


FIG. 1. Sagittal (*left*), axial (*center*), and coronal (*right*) MR images obtained preoperatively showing a large suprasellar arachnoid cyst. Substantial compression on the fornix, corpus callosum, pons, and mesencephalon has caused prominently enlarged lateral ventricles.

1). Both endocrine and ophthalmological examinations were nondiagnostic.

Operation. A neuronavigational MR imaging procedure was performed prior to surgery. We used the Medtronic Stealth navigation system (Medtronic, Inc., Minneapolis, MN) with a tracker attached to a rigid endoscope (MINOP system; B. Braun Aesculap, Tuttlingen, Germany) to make the tip of the scope navigable. The boy's head was fixed in a Mayfield clamp and the cyst was approached through a right coronal bur hole via a transventricular route. Fenestration of the upper cyst wall was performed using the carbon-coated laser tip described by Vandertop, et al.,³² resulting in partial collapse of the cyst. The basal cyst wall proved to be quite thick, which caused reduced visibility of anatomical structures lying caudal to the cyst. The prepontine cistern was localized using neuronavigation, thus allowing fenestration of the lower cyst wall by using this special carbon-coated laser tip as well as scissors. Finally, the membrane of Lilliequist was opened with this laser and scissors, providing a wide communication between ventricles, cyst, and prepontine cistern (Fig. 2).

Postoperative Course. After the surgery, the head bobbing gradually disappeared. The boy's head circumference at the age of 3 years was 51.5 cm (below the 75th percentile). At the 18-month follow-up examination, the abnormal head movements were still completely absent. Neurological and psychological examinations revealed normal motor development and intelligence. Postoperative MR images at the 2-month follow up (Fig. 3) showed a significant decrease in the size of the suprasellar cyst, a diminished mass effect of the cyst, and a reduced volume of the lateral ventricles.

Discussion

Several pathophysiological mechanisms causing BHDS have been described in the literature, including extrapyramidal² or thalamic^{28,29} dysfunction, disturbed function of the nucleus ruber,^{7,17} and learned behavior reducing obstruction of the foramen of Monro by the dome of the cyst.³⁴ Recent-

ly, Hagebeuk and colleagues¹⁷ reviewed the English-language literature and discussed 30 cases of BHDS, as well as the pathophysiological mechanism of head bobbing. Compression of the nucleus ruber and damage in its connections with cerebellothalamic, cerebelloolivary, and nigrostriatal fibers may be implicated in the pathophysiology of the disorder.¹⁷

Although its pathogenesis is still unclear, BHDS can be treated successfully. A number of treatment modalities for suprasellar arachnoid or third ventricular cysts have been suggested, including shunt insertion (ventriculoperitoneal, ventriculoatrial, lumboperitoneal, cystoventriculoperitone-

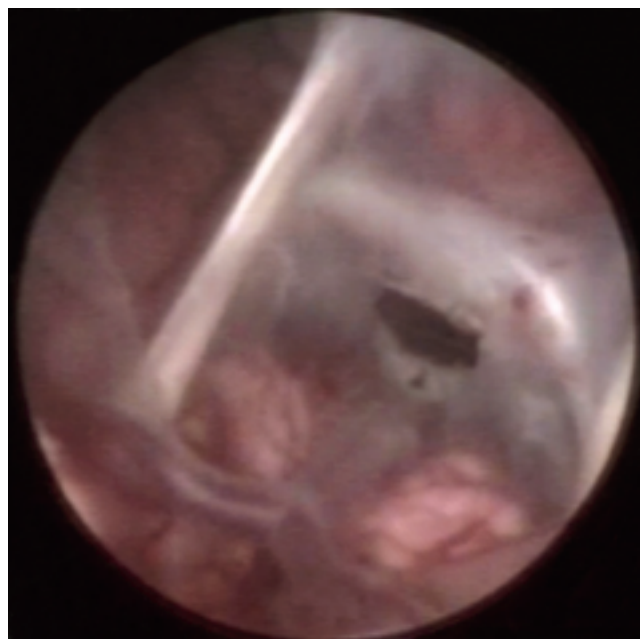


FIG. 2. Endoscopic view showing a large fenestration in the lower cyst wall and the membrane of Lilliequist revealing the prepontine cistern and the pons, basilar artery, and oculomotor nerve.

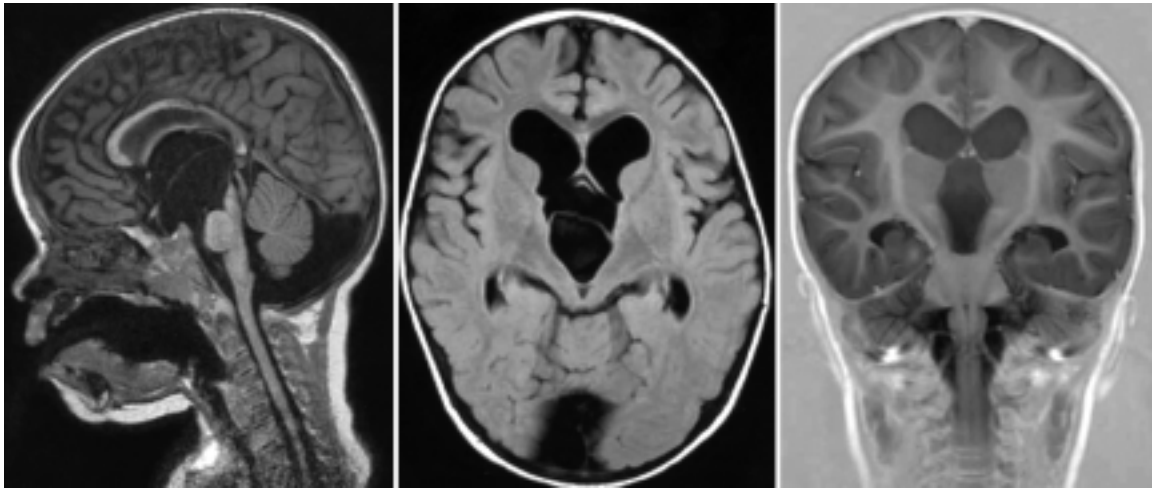


FIG. 3. Sagittal (*left*), axial (*center*), and coronal (*right*) MR images obtained 2 months postoperatively showing a significantly decreased suprasellar arachnoid cyst and mass effect of the cyst, along with reduction of the hydrocephalus.

al, or cystoventriculoatrial) as well as cyst fenestration or resection through craniotomy, endoscopic ventriculocystocisternostomy, or even a combination of techniques.^{1-3,9,10,12,14,20,21,23,24,34,36}

The type of BHDS arising from a third ventricular cyst can be treated with an open (partial) resection or fenestration of the cyst after a right frontal transcortical or transcallosal approach, which results in completely resolved or reduced head bobbing without recurrence of these cysts.^{1,2}

Laser-assisted endoscopic cyst fenestration has been performed with varying success; mostly it results in partial or temporary disappearance of the head bobbing.^{12,20} Many surgeons are not comfortable using endoscopic laser probes in the proximity of vital structures (for example, the basilar artery in the third ventriculostomy). Therefore, in our case a special pretreated laser catheter with a layer of carbon particles was used to fenestrate the lower cyst wall safely. With this pretreated laser catheter tip (“black tip”), the prepontine cistern can be reached safely through careful stepwise perforation of the membranes, even when the anatomy is abnormal or when a thick and multilayered ventricular floor obscures the underlying critical structures.³²

To avoid the placement of a shunt, a ventriculocystocisternostomy was performed in our patient to establish communication between the subarachnoid space and the suprasellar cyst via a minimally invasive procedure.^{6,8,9,26,30} Although a single large fenestration of the cyst wall can be sufficient,¹³ it may be wise to make a second basal fenestration when this is safe, because obliteration of fenestrations is common due to the low flow aspects in these cysts.^{4,5} In this additional step, a fenestration in the lower wall of the cyst is made in an attempt to prevent closure of the ventriculocystostomy.^{5,8} Long-term follow-up MR imaging studies have provided evidence of secondary closure of the upper fenestration, but when a ventriculocystocisternostomy is performed, the basal fenestration remains functional.⁸

The effect of surgical intervention seems to depend on the time elapsed between the onset of the head bobbing and the procedure. Hagebeuk, et al.,¹⁷ confirmed the findings of Mussel, et al.,²⁰ and reported earlier intervention in 20

children with completely resolved head bobbing compared with 10 children in whom head bobbing was only decreased (the mean time from onset of symptoms to surgery was 21 and 63 months, respectively). The mental retardation in their own case of BHDS could be related to the treatment delay of more than 3 years.¹⁷ In our case, the head bobbing completely disappeared, and the boy’s head circumference decreased below the 75th percentile. Additionally, neurological and psychological examinations revealed normal motor development and normal intelligence levels; thus our case confirms the importance of adequate surgical treatment at an early stage.

Combined stereotaxy using neuronavigation and endoscopic third ventriculostomy has been described by several authors.^{11,15,16,27,30,37} Reports of stereotactic endoscopy for the treatment of arachnoid cysts have been published as well.^{18,26,31,35} Schroeder and associates³¹ described the use of a frameless neuronavigation system in six cystocisternostomies in arachnoid cysts. Neuronavigation can be useful during endoscopic procedures for selection of the best entry point into cysts, for the determination of the optimal approach for fenestration in a ventriculocystostomy,^{15,31,37} and for cysts having a thick wall and no visible anatomical references below the basal cyst wall. The combination of image guidance with neuroendoscopic techniques can shorten the operating time by immediate selection of the optimal route and location of the fenestration site, thereby significantly reducing morbidity and mortality rates.^{27,31,33} Problems with brain shift have not been encountered during frameless-neuronavigated ventriculostomy because of continuous irrigation¹⁶ and because the target region is near a rigid structure.

To our knowledge, this is the first reported case of a young child with BHDS due to a large suprasellar arachnoid cyst that was successfully treated using laser-assisted endoscopic ventriculocystocisternostomy and neuronavigation. Our case demonstrates the value of neuronavigation in a laser-assisted endoscopic ventriculocystocisternostomy of a large suprasellar arachnoid cyst. Neuronavigation systems enable orientation during endoscopic procedures when anatomical landmarks are distorted or not visible.

Conclusions

This case report emphasizes the safety of laser-assisted endoscopic ventriculocystocisternostomy, the valuable guidance of neuronavigation, and the importance of early treatment of BHDS. Navigated laser-assisted ventriculocystocisternostomy, performed within 2 years after the onset of symptoms, led to complete cessation of head bobbing, along with normalization of motor development and reduction of the cyst and ventricle sizes on postoperative MR imaging. Therefore, we propose early navigated laser-assisted ventriculocystocisternostomy as a safe, effective, and minimally invasive treatment option for BHDS due to a suprasellar cyst.

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