Bobble-head doll syndrome: report of 2 cases and a review of the literature, with video documentation of the clinical phenomenon

Bryan Renne, PhD,¹ Stefan Rueckriegel, MD,² Sudheesh Ramachandran, MCh,¹
Julia Radic, MD, MPH, FRCSC,¹ Paul Steinbok, MBBS, FRCSC,¹ and
Ash Singhal, MD, MSc, FRCSC¹

¹Faculty of Medicine and the Division of Neurosurgery, University of British Columbia and British Columbia Children's Hospital, Vancouver, British Columbia, Canada; and ²Department of Neurosurgery, University Hospital Würzburg, Germany

Bobble-head doll syndrome (BHDS) is a rare pediatric movement disorder presenting with involuntary 2- to 3-Hz head movements. Common signs and symptoms also found on presentation include macrocephaly, ataxia, developmental delay, optic disc pallor or atrophy, hyperreflexia, tremor, obesity, endocrinopathy, visual disturbance or impairment, headache, and vomiting, among others. The syndrome is associated with suprasellar cysts, third ventricular cysts, or aqueductal obstruction, along with a few other less common conditions. The cause of involuntary head motions is not understood. Treatment is surgical. The authors present 2 cases of BHDS. The first is a 14-year-old boy with BHDS associated with aqueductal obstruction and triventricular hydrocephalus secondary to a tectal tumor. He was successfully treated by endoscopic third ventriculostomy, and all symptoms resolved immediately in the recovery room. This case is unusual in its late age of symptom onset, the primacy of lateral (“no-no”) involuntary head rotations, and the associated tectal tumor. The second case is a 7.5-year-old girl with BHDS associated with a suprasellar cyst. She was successfully treated with an endoscopic fenestration but preexisting endocrinopathy persisted, and the patient was diagnosed with autism spectrum disorder at age 12 years. This second case is more typical of BHDS. A comprehensive and up-to-date review of the literature of BHDS and video documentation of the phenomenon are presented.

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KEY WORDS bobble-head doll syndrome; head bobbing; aqueductal obstruction; tectal tumor; suprasellar cyst; oncology

BOBBLE-HEAD doll syndrome (BHDS) is a rare movement disorder first identified in the literature by Benton et al.⁵ The cardinal feature of BHDS is an involuntary 2- to 3-Hz movement of the head in the anterior-posterior direction (as if nodding “yes-yes” like the “bobbing” of a bobble-head doll toy), in the lateral rotational direction (as if shaking the head “no-no”), or in some irregular alternating combination of the 2 directions.

We present 2 new cases of BHDS. The first has several unusual features, perhaps the most striking being the late age of symptom onset, a greater tendency toward the rarer lateral (“no-no”) motions, and the patient’s inability to voluntarily suppress the motions. The second is a more typical case, although the age of symptom onset is also unusually late.

A comprehensive review of the literature was per-

formed by the first author (B.R.). A search for the term “bobble head” in both PubMed and MEDLINE was carried out. The cases from these reports were combined with those found in previous reviews.^{17,19,23,28,33,44,48,49} Various cases were then excluded: those lacking the cardinal head movement feature^{4,38,41} and those originally reported in publications (often non-English language) that we could not obtain. This yielded a total of 70 previous cases of BHDS. The list of cases extracted from the literature was not confirmed by any of the authors except first author. The list of previously published cases of BHDS combined with the 2 current cases, as well as a summary of their features, is compiled in Table 1.

The most recent BHDS review was published in 2005.¹⁹ This current updated review includes reports published since that time as well as other cases found in the literature (at times part of other series). Thus, in addition to pre-

ABBREVIATIONS BHDS = bobble-head doll syndrome; CVP = cystoventriculoperitoneal; ETV = endoscopic third ventriculostomy; VP = ventriculoperitoneal.

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senting a new atypical case and another more typical one, we provide a comprehensive and up-to-date summary of BHDS as of the time of writing. We also obtained video documentation of the phenomenon, which will provide the clinician with an immediate impression of the clinical features of this condition.

Case Reports

Case 1

History and Presentation

A 14-year-old boy presented with a 3-year history of headache with intermittent vomiting, recent cognitive decline, and a 6-week history of repetitive involuntary motions of the head and trunk that had increased in the past 3 days. Primary motion of the head was in a 2- to 3-Hz lateral-to-lateral rotation (as if shaking his head “no-no”), although some occasional anterior-to-posterior movements (as if nodding “yes-yes”) were intermittently present as well. In addition, the patient would involuntarily flex and extend his trunk from time to time as if attempting to find a comfortable position in the bed. Involuntary head motions would stop whenever the patient would rest the posterior aspect of his head on the bed. Video 1 demonstrates head and truncal motions and gaze testing both before and after surgery.

VIDEO 1. Case 1 was a 14-year-old boy with BHDS associated with a tectal mass. Primary head motions show a 2- to 3-Hz lateral-to-lateral (“no-no”) rotation with occasional anterior-to-posterior (“yes-yes”) movements. Involuntary truncal flexion and extension are present as well. Head motions stop when the patient rests the posterior aspect of his head on the bed. Eye movement testing shows some hesitation in upward gaze. Postsurgical excerpts show absence of involuntary motions and full eye movements. Copyright Bryan Renne. Published with permission. Click here to view.

Examination

On examination, the patient was conscious but slightly confused. He had macrocephaly (60.5-cm head circumference, which is more than 2 standard deviations above the age-sex mean) and pale optic discs. He was not able to stand or walk, so an assessment of gait was not feasible. He did show some lag in upward gaze but otherwise had no neurological deficits other than the repetitive motions of the head and trunk. An MRI study of the head showed a tectal mass causing aqueductal obstruction and obstructive triventriculomegaly (Fig. 1).

Operation and Postoperative Course

An endoscopic third ventriculostomy (ETV) was performed without complications. The patient's head and truncal motions resolved completely in the recovery room immediately after surgery. On 4-month follow-up, the motions had not returned. An MRI study obtained 9 months after surgery did show mild increased size of the tumor; however, the ventricles had decreased in size. Endocrinological follow-up has been unremarkable, though an adrenocorticotrophic hormone (ACTH) stimulation test is scheduled. The patient will be followed up further by endocrinology and oncology departments.

Case 2

History and Presentation

A 7.5-year-old, left-handed girl presented with a 2-year history of recently worsening involuntary anterior-posterior bobbing motions of the head at approximately 3 Hz. The movements were more severe when she was emotionally upset, would stop during sleep, and could be voluntarily suppressed for a couple of minutes. There was no discomfort or exhaustion associated with the head bobbing. A history of headache or vomiting was denied.

Examination

On examination, the patient was alert, oriented, and appeared healthy. She had normal cognitive development. Her head circumference was 53.5 cm (90th percentile). She had short stature (110 cm, which is more than 3 standard deviations below the age-sex mean). Eye movements were full, and there were no visual field deficits. However, bilateral optic atrophy was noted. Motor examination showed Grade 4+ proximal weakness of the upper and lower limbs, with normal strength distally. Reflexes were brisk, more so in the lower limbs (Grade 3+) than in the upper limbs (Grade 2+). Tandem gait was slightly unsteady both backward and forward. Plantar responses were normal, and clonus was absent. The sensory examination showed normal findings. An endocrine assessment revealed normal thyroid, cortisol, and antidiuretic hormone reserve; however, the patient did have growth hormone deficiency. She had a longstanding history of polyuria and polydipsia, but diabetes insipidus was ruled out. An MRI study of the head showed a large suprasellar cyst causing obstructive hydrocephalus (Fig. 2 and Video 2).

VIDEO 2. Case 2 was a 7.5-year-old girl with BHDS associated with a suprasellar cyst. Primary head motions are a 3-Hz anterior-to-posterior (“yes-yes”) movements. Gait testing is demonstrated. Intraoperative excerpt shows cyst fenestration and cautery. Postsurgical excerpt shows absence of involuntary head motions. Copyright Bryan Renne and Paul Steinbok. Published with permission. Click here to view.

Operation and Postoperative Course

An endoscopic fenestration of the cyst was performed. Cisternostomy was attempted but was aborted due to the occurrence of bradycardia. Because the cisternostomy could not be performed, the cyst wall was shrunk using cautery. The patient's head bobbing resolved in the recovery room immediately after surgery. However, intermittent anterior-posterior bobbing did return on the 2nd postoperative day, although it was greatly diminished compared with that seen at initial presentation and occurred only during times of fatigue or emotional upset. This intermittent head bobbing eventually ceased as the patient was followed up over a 7.5-year period. The patient was started on growth hormone shortly after surgery. Her growth over the follow-up period put her not far from the height expected based on her parents' heights. The patient was diagnosed at age 12 with autism spectrum disorder.

Discussion

The cardinal feature of BHDS is an involuntary 2- to

TABLE 1. Two BHDS cases currently presented and summary of those reported in the literature

Authors & Year	Age (yrs)		Sex	Diagnosis	Non-HB Signs & Symptoms	1° HB	Can Stop HB?*	Tx	Post-Tx HB (complications)
	Onset	Tx							
Benton et al., 1966	2.00	3.25	F	3VC	Ataxia, macrocephaly, intell delay, hyperreflexia, optic disc pallor/atrophy, tremor, visual dist/impairment, headache, tactile hyperesthesia, right Babinski sign	V	Y	Excision; 1 yr later, VP shunt (for hydrocephalus)	Gone w/in 1 yr (diabetes insipidus, hypothyroidism, obesity, hydrocephalus, hypertonicity)
Nelhaus, 1967	3.50	6.50	F	3VC	Macrocephaly, hyperreflexia, optic disc pallor/atrophy, tremor, obesity, alternating strabismus, impaired visuomotor perception	V	Y	Fenestration	Less
	9.00	10.00	M	AO	Ataxia, macrocephaly, motor delay, hypertonicity, hyperreflexia, mild esophoria, impaired visuomotor perception	H	Y	Ventriculostomy, Pudenz valve; 6 wks later, ventricular drainage, removal of Pudenz valve, insertion of Holter valve (for bilat subdural hematoma)	Gone w/in 8 mos (bilat subdural hematoma)
	3.00	6.00	M	3V mass	Macrocephaly	V	—	None (HB only while walking)	Same
Nishiura, 1969	7.50	16.00	M	Chronic hydrocephalus	Ataxia, intell delay, hyperreflexia, optic disc pallor/atrophy, visual dist/impairment, obesity, behavioral disorder, suicidal ideation, psychopathology	V	Y	Ventriculostomy, Pudenz valve	Less
Mayher & Gindin, 1970	0.58	2.50	M	3VC	Macrocephaly, motor delay, weakness, hyperreflexia, optic disc pallor/atrophy, teeth grinding	V	S	Excision	Gone (hemiparesis, seizures, death)
Sakata et al., 1971	—	13.00	M	SC	HB only while walking	B	—	Fenestration, VP shunt	Less
Obenchain & Becker, 1972	3.00	26.00	F	3VC	Macrocephaly, hyperreflexia, optic disc pallor/atrophy, visual dist/impairment, headache, episodic loss of consciousness	V	Y	Excision; 9 days later, ventriculogular shunt (for headaches, increased ICP)	Less
Patriquin, 1973	3.50	6.00	M	3VC	Ataxia, macrocephaly, motor delay, weakness, headache, vomiting, obesity, Gower's sign	V	Y	Excision	Gone w/in 10 days
Ferrey et al., 1974	2.50	5.00	M	SC	Ataxia, macrocephaly, optic disc pallor/atrophy, rt local motor seizure	V	Y	Aspiration & drainage	Gone
Russo & Kindt, 1974	0.42	1.00	F	AO	Macrocephaly, devel delay, hypertonicity, hyperreflexia, lethargy	V	Y	VA shunt	Gone after 8 days
Segall et al., 1974	1.08	3.25	M	SC	Ataxia, optic disc pallor/atrophy, tremor, precocious puberty	V	Y	Excision	Gone
Görke et al., 1975	1.67	7.00	M	SP cyst	Motor delay, intell delay, weakness, hypotonicity, hyperreflexia, Gower's sign	V	Y	Fenestration	Less: after 6 mos, present only when emotionally excited (precocious puberty)

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Authors & Year	Age (yrs)			Diagnosis	Non-HB Signs & Symptoms	1° HB	Can Stop HB?*	Tx	Post-Tx HB (complications)
	Onset	Tx	Sex						
Russman et al., 1975	2.50	9.00	M	CVI cyst	Macrocephaly, tremor, hyperesthesia to warm water (at 4.5 yrs; resolved by 9 yrs old), bilat dysdiadochokinesia, declining school performance	V	Y	VA shunt	Less after 4 mos
	5.50	10.00	M	SC	Macrocephaly, hyperreflexia, tremor, disturbed behavior, suicidal ideation	H	Y	Incision & drainage; 3 wks later, VA shunt (for diabetes insipidus, headaches)	Less: after 5 mos, present only w/ anxiety or walking (diabetes insipidus & headaches resolved w/ shunt)
Tomasovic et al., 1975	0.33	0.83	F	AO	Ataxia, macrocephaly, motor delay, hyperreflexia, EMD, bilat ankle clonus	V	S	VP shunt	Less w/in 4 mos
	4.50	5.50	F	AO	Ataxia, macrocephaly, motor delay, language delay, tremor	V	Y	VP shunt	Gone after several days
Deonna & Dubey, 1976	1.00	1.58	F	AO	Macrocephaly, motor delay	H	S/P	Drainage via Torkildsen procedure	Gone
Kirkham, 1977	0.75	4.50	M	3VC	Ataxia, optic disc pallor/atrophy	V	—	Laceration; 10 mos later, VP shunt (for vomiting, headaches, lethargy, hydrocephalus)	Gone, then returned after 1 mo; after shunting, gone w/ in 2 yrs
	3.50	13.00	M	3VC	Ataxia, obesity	V	Y	Excision	Gone after 2 wks
Jensen et al., 1978	0.67	5.00	M	3VC	Ataxia, hypotonicity, obesity	V	Y	Excision	Less after 1 wk
	—	0.92	—	AO	—	—	—	Shunt	Same
	—	1.50	—	SC	—	—	—	Shunt	Same
	—	9.00	—	3VC	—	—	—	Excision	Less
	—	2.00	—	SC	—	—	—	None	Same
	—	13.00	—	SC	—	—	—	Fenestration, shunt	Less
	—	12.00	—	Craniopharyngioma	—	—	—	—	—
	—	—	—	SC	—	—	—	—	—
	—	—	—	SC	—	—	—	—	—
Albright, 1981	9.50	9.50	M	3VC	Macrocephaly, intell delay, hyperreflexia, optic disc pallor/atrophy, obesity, memory loss, precocious puberty, short stature	V	Y	Excision; had previous VP shunt age 6 yrs (for ataxia), revised age 9 yrs (for nausea, vomiting, headache)	Less
Dell, 1981	—	11.00	F	3VC	Ataxia, intell delay, optic disc pallor/atrophy, EMD, chronic seizure disorder, graying of hair	V	—	Fenestration, drainage	Gone (fevers that resolved spontaneously after multiple courses of antibiotics)
	0.42	0.50	—	Shunt malfunction	Motor delay, EMD, meningocele L2-L3 to sacrum, preexisting VP shunt (for increasing head circumference), hyporeflexia, absent ankle jerks	V	P	Shunt revision; then 3 shunt revisions over 8 mos (for shunt malfunctions)	Gone w/in 8 mos of initial revision

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Authors & Year	Age (yrs)		Sex	Diagnosis	Non-HB Signs & Symptoms	1° HB	Can Stop HB?*	Tx	Post-Tx HB (complications)
	Onset	Tx							
Kaplan et al., 1984	1.17	2.00	F	SC	Macrocephaly, optic disc pallor/atrophy	H	Y	Endoscopic fenestration, VP shunt; 3 wks later, CVP shunt (for HB recurrence)	Gone, then returned after 3 wks; after CVP shunt, gone w/in 1 yr
Wiese et al., 1985	7.00	13.00	F	SC	Macrocephaly, optic disc pallor/atrophy, visual dist/impairment, headache, obesity, intermittent hyperhidrosis but rt-sided anhidrosis, lt lower-extremity hyperalgesia to pinprick below L-1	V	Y	LP shunt	Less after 5 mos
Harsh et al., 1986	—	7.00	F	SC	Devel delay	—	—	Fenestration	Less (transient fever & lethargy)
Coker, 1986	0.50	0.50	M	Trapped 4V (no enlarged 3V)	Preexisting shunt at age 4 mos (for lethargy & "setting sun" eye sign sec to AO & fever presumed sec to meningitis), dilation of 4th ventricle & aqueduct at age 6 mos	V	—	Shunt; 2 mos later, revised (4V dilation, arachnoiditis)	Gone, then returned after 2 mos, then gone after revision
Notholt-Heerich et al., 1987	3.50	4.10	F	3VC	Ataxia, macrocephaly, motor delay, optic disc pallor/atrophy	V	D	VA shunt, Ommaya reservoir; 6 mos later, CVP shunt (for HB recurrence)	Gone, then returned after 6 mos, then less after CVP shunt
Parizek et al., 1989	3.00	4.50	F	3VC	Ataxia, optic disc pallor/atrophy, tremor, mild left-sided hemiparesis	V	—	2 CVP shunts; 2 days later, 1 replaced (for misplacement)	Gone after 3 mos (precocious puberty)
	0.33	10.00	F	3VC	Tremor, vomiting, diabetes insipidus, bulimia, sleeplessness, enuresis	V	—	CVP shunt	Gone after 3 wks (evacuation of pseudocyst at end of catheter after 4.5 yrs)
	3.00	14.00	M	3VC	devel delay, diabetes insipidus, bilat amaurosis, restlessness, confabulation, aggressiveness	V	—	Drainage via bilat Torkildsen procedure; 3 yrs later, CVA shunt	Same; after CVA shunt, gone w/in 3 mos
Turgut & Özcan, 1992	6.00	8.00	M	3VC	Intell delay, precocious puberty	V	—	Excision	Gone after 6 mos
Pollack et al., 1995	1.75	2.00	M	Cystic choroid plexus papilloma	Episodic rt anterolat head tilt; drop attacks progressing in hours to coma	V	Y	Ventriculostomy, endoscopic fenestration SP; 4 days later, resection	Gone after resection
Ahn et al., 1997	10.00	11.00	M	Misplacement catheter tip of shunt	Ataxia, intell delay, language delay, hyperreflexia, headache, vomiting, attention deficit disorder, right esotropia	H	N	Revision (of SDP shunt placed 3 yrs earlier for noncommunicating subdural fluid collection in rt cerebral hemisphere)	Gone
Mussell et al., 1997	1.00	5.50	F	SC	Ataxia, macrocephaly, hyperreflexia, optic disc pallor/atrophy, tremor, visual dist/impairment, vomiting, obesity, precocious puberty, urinary incontinence, ankle clonus	V	Y	Endoscopic fenestration; 3 mos later, VP shunt (for headache, vomiting, incontinence, worsening HB)	Less; 6 mos after shunt, HB visible only when walking
Goikhamn et al., 1998	—	2.17	F	3VC	Ataxia, macrocephaly, motor delay, hypotonicity, hyperreflexia	V	Y	Excision, CVP shunt	Gone w/in days

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Authors & Year	Age (yrs)		Sex	Diagnosis	Non-HB Signs & Symptoms	1° HB	Can Stop HB?*	Tx	Post-Tx HB (complications)
	Onset	Tx							
Sharma et al., 2001	14.00	14.00	F	Trapped 4V (sec to lateral ventricular shunting)	Ataxia, headache, vomiting, bilat abducens palsies, neck stiffness	V	S/P	4V VP shunt (preexisting lateral ventricular VP shunt, revised twice, for congenital hydrocephalus due to AO)	Gone w/in 1 wk
Bhattacharyya et al., 2003	12.00	13.00	M	AO (web)	Ataxia, macrocephaly, hyperreflexia, resuscitated from coma (presumed brainstem herniation) 1 wk earlier	V	Y	Dehydration therapy (due to coma mid-workup); 5 days later, when back to baseline, VP shunt for AO	Gone
Desai et al., 2003	—	2.00	F	SC	—	V	Y	Marsupialization	Gone
	—	3.00	F	SC	—	V	Y	Marsupialization	Gone
	—	5.00	F	SC	—	V	Y	Marsupialization	Gone
Fioravanti et al., 2004	—	1.00	F	SC	Persistent nocturnal tearing, previous VP shunt (for supratentorial hydrocephalus, still unresolved)	—	S	Endoscopic coagulation, fenestration; 18 mos later, endoscopic drainage (for HB recurrence)	Less; after 18 mos, returned; after drainage, gone w/ in 3 yrs
	—	9.00	M	SC	Ataxia, precocious puberty	—	S	Endoscopic fenestration	Gone
Hagebeuk et al., 2005	0.25	4.00	M	SC	Ataxia, inability to jump	V	Y	Endoscopic fenestration, balloon dilation of communication w/ basal preoptine cistern	Gone w/in 3 years
Zamponi et al., 2005	1.00	3.00	—	3VC, SP cyst	Ataxia, tremor	V	S	VP shunt	Gone
Van Benijum et al., 2006	1.00	2.00	M	SC	Macrocephaly, motor delay	V	S	Endoscopic fenestration	Gone w/in 18 mos
de Brito Henriques et al., 2007	1.00	1.00	F	Dandy-Walker syndrome (no 4V hypertension)	Posterior fossa cyst, cerebellar vermis hypoplasia	H	D/S	2 suboccipital punctures testing for hypertension in 4V, then conservative management; had previous VP shunt day 9 of life (for Dandy-Walker syndrome)	Same
Guerreiro et al., 2012	9.00	10.00	F	SC	Hyperreflexia, nocturnal hunger & enuresis, 13-kg weight gain, impaired short-term memory & attention	V	N	Endoscopic ventriculocystostomy; 3 mos later, CVP shunt (for HB recurrence)	Gone, then returned after 3 mos; after CVP shunt, less, & then gone by age 16 yrs
	3.00	11.00	M	SC	Macrocephaly, obesity	V	—	Endoscopic ventriculocystostomy	Gone, then returned after 3 mos; lost to follow-up
	0.83	0.92	M	SC	Ataxia, macrocephaly, devel delay, tremor, tone disturbance	V	—	Ventriculocystostomy	Gone w/in 1 wk
	0.75	1.00	M	SC	Macrocephaly, devel delay, hyperreflexia, optic disc pallor/atrophy, visual dist/impairment	V	—	VP shunt (to treat hydrocephalus); 17 mos later, endoscopic ventriculocystostomy & removal of VP shunt	Gone then returned after 3 mo; after endoscopic procedure, gone

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TABLE 1. Two BHDS cases currently presented and summary of those reported in the literature

Authors & Year	Age (yrs)		Sex	Diagnosis	Non-HB Signs & Symptoms	1° HB	Can Stop HB?*	Tx	Post-Tx HB (complications)
	Onset	Tx							
Garg et al., 2012	20.00	20.00	M	VP shunt drainage malfunction (status post-Tx for tuberculous meningitis)	Optic disc pallor/atrophy, visual dist/impairment	B	S	None (HB began 24 hrs after VP shunt for ventriculomegaly status 1 mo post-Tx for tuberculous meningitis)	Same; lost to follow-up
Ishihara et al., 2013	0.25	0.33	M	PF cyst (causing hydrocephalus)	Macrocephaly	H	S	None (no 3V dilation; had previous endoscopic fenestration day 12 of life for congenital PF cyst causing hydrocephalus)	Almost gone by age 1 yr
Alexiou et al., 2013	4.00	6.00	M	SC	—	V	N	CVP shunt	Gone
Sridhar et al., 2013	6.00	9.00	M	SC	Devel delay, poor height gain, aggression, tactile hallucinations	—	S	Endoscopic fenestration, cautery	Gone
Reddy et al., 2014	8.00	9.00	M	SC	Optic disc pallor/atrophy	V	Y	Endoscopic fenestration	Less
Bhattacharyya et al., 2014	11.00	11.00	M	AO (pineal mass)	Ataxia, optic disc pallor/atrophy, visual dist/impairment, papilledema, sluggish direct & consensual responses, bilateral relative afferent pupillary defect	B	Y	VP shunt	Gone
Mala et al., 2014	3.00	4.00	M	SC	—	V	S	Ventriculocystostomy	Unknown (died on postop day 2 w/ seizures)
Ramesh & Raju, 2015	0.08	0.83	F	SC	2 tonic-clonic seizures	—	—	Endoscopic excision, marsupialization, 3rd ventriculostomy, septostomy	Gone
	8.00	8.00	F	SC	Ataxia, intermittent fever, 1 tonic-clonic seizure	V	N	Endoscopic ventriculocystostomy, 3rd ventriculostomy, reservoir	Less
	1.00	4.00	F	SC	—	V	S	Endoscopic marsupialization, ventriculocystostomy, 3rd ventriculostomy	Gone
Present report, Case 1	14.00	14.00	M	AO (tectal tumor)	Macrocephaly, intell delay, optic disc pallor/atrophy, vomiting, lag in upward gaze	H	P	Endoscopic 3rd ventriculostomy	Gone
Present report, Case 2	5.50	7.50	F	SC	Hyperreflexia, optic disc pallor/atrophy, upper- & lower-limb proximal weakness, growth hormone deficiency, longstanding polyuria & polydipsia (no diabetes insipidus)	V	Y	Endoscopic fenestration, cautery	Gone; occasional HB returned after 2 days, then gone after 7.5 yrs

AO = aqueductal obstruction; B = both horizontal and vertical; CVI = cavum velum interpositum; CVP = cystoventriculoperitoneal; D = head motions stop with distraction; devel = developmental; dist = disturbance; EMD = eye movement disorder; H = horizontal (lateral); HB = head bobbing (involuntary head motions); ICP = intracranial pressure; intell = intellectual; N = head motions cannot be stopped voluntarily; P = head motions stop with supine position; PF = posterior fossa; S = head motions stop with sleep; SC = suprasellar cyst; SDP = subdural peritoneal; sec to = secondary to; SP = septum pellucidum; Tx = treatment; V = vertical (anterior-posterior); VA = ventriculoatrial; VP = ventriculoperitoneal; Y = head motions can be stopped voluntarily; 1° HB = primary direction of head bobbing (involuntary head motions); 3V = 3rd ventricle; 3VC = 3rd ventricular cyst; 4V = 4th ventricle; — = unknown.

* In this column only, a forward slash ("/") indicates that there were multiple reported ways involuntary head motions could be stopped (e.g., "SP" indicates motions can be stopped with sleep and with being in the supine position); however, if motions could be voluntarily suppressed, only a "Y" was recorded.

FIG. 1. Case 1. MR images obtained in a 14-year-old boy with BHDS associated with a tectal mass (white and black arrows). **A:** Presurgical axial T2-weighted image showing obstructive triventriculomegaly. **B:** Post-ETV axial T2-weighted image showing reduced size of the third and lateral ventricles. **C:** Presurgical sagittal turbo inversion recovery magnitude image showing aqueductal obstruction by tectal tumor causing triventricular dilation, bowing of the lamina terminalis and the floor of the third ventricle, and sellar erosion. **D:** Post-ETV sagittal magnetization-prepared rapid gradient-echo image showing a relaxed floor of the third ventricle. Post-ETV images were taken 9 months after the presurgical images and showed that the tectal mass increased in size to $19 \times 25 \times 24 \text{ mm}^3$ (craniocaudal $\times$ anteroposterior $\times$ right-left) from a presurgical size of $19 \times 20 \times 20 \text{ mm}^3$.

3-Hz movement of the head in the anterior-posterior (“yes-yes”) direction, the lateral (“no-no”) rotational direction, or some irregular alternating combination of the 2 movements. In addition to involuntary head and truncal motions, information on presenting signs and symptoms was available in 58 of the 72 cases (81% of total, see Table 1). Common presenting signs and symptoms are summarized in Table 2. Other less common signs and symptoms not included in Table 2 are listed with individual cases in Table 1.

The age of onset of abnormal head movements was documented in 55 cases (76% of total), and in these cases the median onset age was 3.0 years (median absolute deviation of 3.35 years), with a range of 1 month to 20 years. Thus, BHDS is typically a disease of early childhood, and the onset age of 14 years in our Case 1 is the second oldest reported in the literature.

Of the 62 cases (86% of total) for which patient sex is identified, 35 were male (56%) and 27 female (44%) for a male/female ratio of 1.3:1. Previous reviews, which relied on a subset of the cases we have identified, report a higher male/female ratio of 2:1.

Of the 59 cases (82% of total) for which there was enough information to identify a direction of repetitive head movements, 48 were primarily anterior-posterior (i.e., 81% “yes-yes”) and 8 were primarily lateral (i.e., 14% “no-no”). For patients with both anterior-posterior bob-

FIG. 2. Case 2. MR images acquired in a 7.5-year-old girl with BHDS associated with suprasellar cyst. Presurgical axial T2-weighted image (**A**) and presurgical sagittal constructive interference in steady-state (CISS) image (**C**) showing a large suprasellar cyst. Post-fenestration axial T2-weighted image (**B**) and post-fenestration CISS image (**D**) showing reduced size of the cyst.

bing (“yes-yes”) and lateral shaking (“no-no”) movements, one direction took primacy over the other in all but 3 cases (5%). The BHDS literature often mentions that head motions decrease or cease with sleep, distraction, or with patients in the supine position; increase with excitement or anxiety; and are voluntarily suppressible for a period of seconds to minutes. Regarding the increase or decrease in motion, the literature is often unclear as to whether this change refers to frequency, amplitude, perceived effort on the part of the patient, or something else. As for whether head motions are suppressible, of 50 cases (69% of the total) in which this information was available, movements were reported as voluntarily suppressible for 29 patients (58%); as not voluntarily suppressible for 4 (8%); and as suppressible with one or more distraction, or sleep, or being in the supine position (and either not voluntarily suppressible or without mention of being voluntarily suppressible) in 17 (34%). Of the 37 patients (51% of total) treated at age 3 years or more with available data on whether head motions were suppressible, 24 patients (65%) could suppress motions voluntarily, while 13 (35%) either could not suppress head motions voluntarily or could do so only with one or more of distraction, or sleep, or being in the supine position. This suggests that repetitive head motions in BHDS can usually be suppressed, and, if the child is at least 3 years old at the time of treatment, suppression is most often voluntary. These considerations highlight 2 other rare features of our case 1: This 14-year-old patient could not suppress his head motions voluntarily (although he could do so when supine), and his primary direction of motion was lateral shaking (“no-no”). Our case 2 is more typical, in that the patient could temporarily suppress her head motions and the primary direction of motion was anterior-posterior bobbing (“yes-yes”).

TABLE 2. Presenting signs and symptoms of BHDS

Signs & Symptoms	No. of Cases (of 58)	% of Cases
Macrocephaly	27	47
Ataxia	25	43
Impairment or developmental delay (cognition, motor, or language)	24	41
Optic disc pallor or atrophy	21	36
Hyperreflexia	18	31
Tremor	11	19
Obesity	9	16
Endocrinopathy (including precocious puberty)	9	16
Visual disturbance or impairment	8	14
Headache	6	10
Vomiting	6	10
Weakness	3	5
Hypotonicity	3	5
Eye movement disorders	3	5
Hypertonicity	2	3

Presenting signs and symptoms other than involuntary head motions were reported in 58 cases (81% of total).

In the 72 cases of this review, the major pathologies associated with BHDS were as follows:

- 32 cases (44%): suprasellar cyst (including our Case 2).^{3,12–14,18–20,23,24,26,28,36–38,40,41,43,47,48}
- 18 cases (25%): third ventricular cyst,^{2,5,10,16,23,25,27,31–34,46,49} in one case concurrent with a septum pellucidum cyst.⁴⁹
- 10 cases (14%): aqueductal obstruction (aqueductal stenosis 8,^{11,23,29,39,44} aqueductal web 1,⁷ pineal mass 1,⁶ and tectal tumor 1 [our case 1]); 1 patient had an associated isolated trapped fourth ventricle.⁸
- 4 cases (6%): shunt malfunction, misplacement, or complication, of which 2 were due to malfunction,^{1,10} 1 was due to a trapped fourth ventricle after lateral ventricular shunting,⁴² and 1 was due to catheter placement obstructing the left foramen of Monro.¹⁵
- 3 cases (4%): other cysts impinging on the third ventricle, of which 1 was a septum pellucidum cyst,¹⁷ 1 was a cyst of the cavum velum interpositum,³⁸ and 1 was a cystic choroid plexus papilloma causing obstructive hydrocephalus.³⁵
- 2 cases (3%): masses impinging on the third ventricle, 1 a third ventricular mass²⁹ and 1 a craniopharyngioma.²³
- 2 cases (3%): after fenestration of posterior fossa cysts, 1 after endoscopic fenestration but without enlargement of the third ventricle²² and 1 in a patient with

Dandy-Walker syndrome who did not have intracranial hypertension at the onset of head bobbing.⁹

- 1 case (1%): chronic hydrocephalus.³⁰

The pathophysiology and causative anatomical substrate of BHDS is unknown, although various mechanisms have been suggested: distortion of structures or pathways adjacent to the third ventricle, perhaps involving the basal ganglia (since motions can often be voluntarily suppressed);^{5,29,30} compression of the dorsomedial nucleus of thalamus or the dentatorubrothalamic pathway;^{19,39} a possible role of fetal viral infection in anterior horn cells;¹⁷ the mammillary-fornical-hippocampal circuit;² a “learned behavior” causing transient relief of foraminal blockage or cystic volume reduction,^{35,48} which was demonstrated in one case using a CT cisternogram;⁴⁸ pressure on the dorso-medial red nucleus;⁸ and cerebellar dysfunction,^{9,22} which has been noted to cause abnormal head motions.^{21,45}

Treatment of BHDS is surgical, with the particular procedure depending on the associated pathology. Generally the treatment involves restoration of CSF flow and reducing pressure on compressed structures.

BHDS symptom recurrence has been reported in 8 cases (see Table 1), including our case 2:

- A patient with third ventricular cyst treated with laceration showed return of head motions within 1 month; a ventriculoperitoneal (VP) shunt ceased motions within 2 years.²⁵
- A patient with a suprasellar cyst treated with endoscopic fenestration and a VP shunt had return of head motions after 3 weeks; a cystoventriculoperitoneal (CVP) shunt ceased motions within 1 year.²⁴
- A patient with trapped fourth ventricle treated by shunt placement had return of head motions after 2 months; these were eliminated after shunt revision.⁸
- A patient with obstructive hydrocephalus secondary to third ventricular cyst treated with a ventriculoatrial shunt and an Ommaya reservoir had head motion recurrence after 6 months; these were lessened but not eliminated after CVP shunt placement.³¹
- A patient with suprasellar cyst treated with endoscopic ventriculocystostomy had head motion recurrence after 3 months; a CVP shunt led to elimination of motions within a 16-year follow-up period.¹⁸
- A patient with a suprasellar cyst treated with endoscopic ventriculocystostomy had head motion recurrence after 3 months; the patient was subsequently lost to follow-up.¹⁸
- A patient with obstructive hydrocephalus secondary to a suprasellar cyst treated with VP shunting had head motion recurrence after 3 months; removal of the shunt and endoscopic ventriculocystostomy eliminated head motions.¹⁸

- In our case 2, an SC was treated with endoscopic fenestration and cautery, and occasional head motions returned after 2 days, but head motions disappeared completely within the 7.5-year follow-up.

Thus, symptom recurrence in BHDS is not often reported (8 [11%] of 72 cases reviewed), has occurred within 6 months of initial treatment in the cases known to us, and does not always require treatment. Nevertheless, given the possibility of growth of mass or recurrence of cyst, we elect to follow our BHDS patients with annual MRI. Assuming successful treatment and stable posttreatment clinical course, we transition to less frequent imaging as time progresses.

In conclusion, we have presented 2 new cases of BHDS and have performed a comprehensive review of the literature. We have also provided video documentation of the phenomenon, which will give the clinician an immediate impression of the clinical features of BHDS.

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Disclosures

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Author Contributions

Conception and design: Renne, Rueckriegel, Steinbok, Singhal. Acquisition of data: all authors. Analysis and interpretation of data: Renne, Rueckriegel, Radic, Steinbok, Singhal. Drafting the article: Renne, Steinbok, Singhal. Critically revising the article: all authors. Reviewed submitted version of manuscript: all authors. Approved the final version of the manuscript on behalf of all authors: Renne. Study supervision: Singhal.

Supplemental Information

Videos

Video 1. <https://vimeo.com/240166777>.

Video 2. <https://vimeo.com/240166907>.

Correspondence

Bryan Renne: BC Children's Hospital, Vancouver, BC, Canada. bryan@renne.org.