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# Bobble Head Doll Syndrome. A series of 3 clinical cases managed at the Yalgado Ouédraogo University Hospital and review of the literature

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## ABSTRACT

**Introduction.** Bobble head doll syndrome is a neurological syndrome characterized by abnormal involuntary movements of the head combining repetitive or episodic movements of 2 to 3 Hz back and forth in the anterior-posterior direction that can be assimilated to approval movements (yes-yes) and occasionally lateral rotations that can be assimilated to disapproval signs (no-no). It is a rare entity first described by Benton[1]. We describe three cases managed at the Yalgado Ouédraogo University Hospital in Burkina Faso.

**Observations.** Three children, one girl and two boys, aged respectively 5, 9 and 14 years were seen for involuntary abnormal head movements associating lateral rotations assimilated to signs of disapproval (no-no) in two cases, and back and forth movements of the head in one case. In the 14-year-old adolescent, there was also a picture of intracranial hypertension. Clinical examination revealed ataxia and macrocrania in both cases and a syndrome in 1 case. Imaging revealed triventricular hydrocephalus on aqueductal stenosis in all cases with an associated supra sellar cyst in 2 cases. The treatment consisted of endoscopic treatment in 2 cases and ventriculoperitoneal shunt in 1 case. Surgery allowed a considerable regression of involuntary movements of the head in the immediate postoperative period and a complete recovery in the long term.

**Conclusion.** The bobble head doll syndrome is a rare entity related to the consequences of chronic hydrocephalus responsible for abnormal movements whose management done well and early leads to favourable results.

## INTRODUCTION

Bobble head doll syndrome is a rare pediatric neurological syndrome characterized by abnormal involuntary head movements with repetitive or episodic 2-3Hz back-and-forth movements in the anterior-posterior direction that can be assimilated to approval movements (yes-yes) and occasional lateral rotations that can be assimilated to

## Keywords

Bobble-Head-Doll-Syndrome, endoscopy, hydrocephalus



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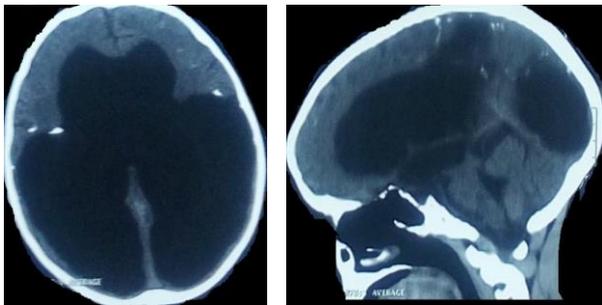
disapproval signs (no-no). This condition was first described in 1966 by Benton[1]. It is a complication of chronic hydrocephalus. The intracranial anomalies that are commonly observed are: non-communicating hydrocephalus, supra sellar arachnoid cyst, septum pellucidum cyst, choroid plexus papilloma, stenosis of the mesencephale aqueduct[5,7,8,10]. We report 03 cases observed and managed at the Yalgado Ouédraogo University Hospital of Burkina Faso with review of the literature.

## CLINICAL ASPECTS

### Case report No. 1

A 9-year-old child, attending school, was admitted for abnormal lateral head movements and recent blindness associated with walking disorders evolving since the age of 3 years. The abnormal movements were initially intermittent and later on permanent, causing, with the onset of blindness, an enormous hindrance to the good progress of schooling. Moreover, this child had a history of two congenital cataract surgeries without favorable results. The physical examination noted normal consciousness, a cranial perimeter of 56 cm, bilateral blindness, microphthalmia, and permanent lateral movements of the head in the form of lateral rotation, which could be assimilated to signs of disapproval (no-no).

Encephalic computed tomography (CT) showed triventricular hydrocephalus by stenosis of the mesencephalic aqueduct with major lamination of the cerebral cortex (Figure 1), and diffuse parenchymal and subarachnoid calcifications.

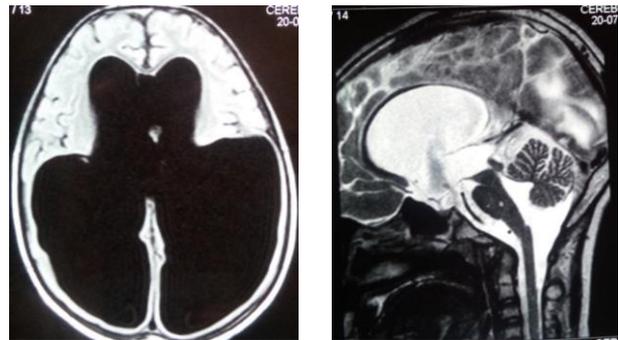


**Figure 1.** Preoperative CT images of case N° 1. **a)** axial section, **b)** sagittal reconstruction showing triventricular hydrocephalus with cortical lamination predominating in the parietal and occipital lobes, and intra parenchymal calcifications.

An indication for ventriculocisternostomy was given and performed.

The postoperative course was initially favorable with considerable regression of the abnormal movements but 4 weeks later, there was a resumption of lateral movements associated with spasms described as electrical discharges and generalized hyperesthesia treated as sensory epilepsy.

Magnetic resonance imaging (MRI) of the brain revealed persistent triventricular hydrocephalus (Figure 2). Therefore, a new ventriculocisternostomy was performed.



**Figure 2.** MRI images of case N° 1 performed 4 weeks after the first ventriculocisternostomy at the resumption of abnormal movements. **a)** T1 axial slice; **b)** T2 sagittal slice showing the persistence of a significant triventricular hydrocephalus.

The clinical evolution was marked by a total regression of the abnormal movements.

Two months later he was readmitted for a left hemiparesis of progressive onset, associated with moderate headaches. A follow-up CT scan revealed a chronic right hemispheric subdural hematoma with a mass effect on the medial structures. The hematoma was evacuated and drained, and the postoperative course was favorable, marked by a complete recovery of the hemiparesis.

Three months later he was readmitted again for a resumption of involuntary head movements; after a cranioencephalic CT scan which showed no other abnormality apart from hydrocephalus, we concluded that the ventriculocisternostomy had failed and indicated a ventriculoperitoneal shunt with a medium pressure valve, which was immediately performed. After this bypass, the abnormal movements disappeared completely and definitively.

### Case report No. 2

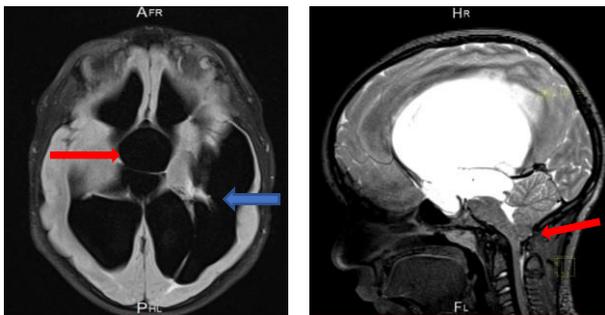
A 14-year-old boy suffering from chronic headaches

was admitted for involuntary head movements associated with a recent worsening of the headaches with the occurrence of vomiting in an intracranial hypertension picture. The history also revealed episodes of generalized seizures and a notion of running away.

On physical examination, we noted a psychomotor slowing down, a head circumference of 58 cm, permanent lateral movements of the head with lateral rotation that could be assimilated to signs of disapproval (no-no). These movements were aggravated by arousal, diminished during voluntary movements and disappeared during sleep. These movements could also be suspended for a few seconds to a few minutes on demand. There was also a spastic right hemiparesis with a motor strength rated at 4/5, a static and locomotor ataxia. Brain MRI (Figure 3) showed tri-ventricular hydrocephalus, a supra sellar cyst exerting a mass effect on V3 and a left temporo-parietal arachnoid cyst. There was also a ptosis of the cerebellar tonsils suggestive of an Arnold Chiari type I malformation.

A ventriculoperitoneal shunt was performed with a medium pressure valve.

The immediate postoperative course was favorable, with improvement of the intracranial hypertension syndrome and significant regression of involuntary head movements. At three months post-op, the abnormal head movements had completely disappeared.



**Figure 3.** Brain MRI of case N° 2. **a):** T1 axial slice; **b):** T2 sagittal slice showing triventricular hydrocephalus with supra sellar cyst (red arrow on axial slice), left temporo-parietal arachnoid cyst (blue arrow on axial slice) and Arnold Chiari I malformation (arrow on sagittal slice).

### Case report No. 3

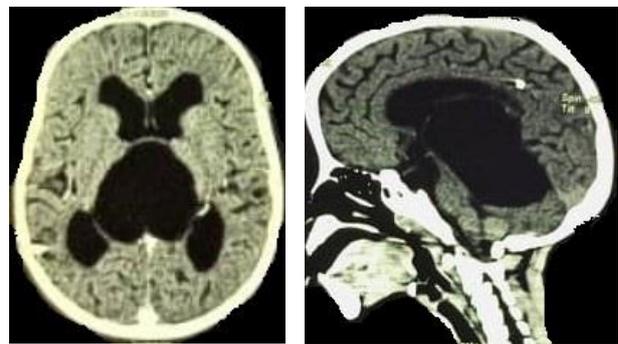
A 4-year-old girl operated at the age of 2 years for triventricular hydrocephalus (ventriculoperitoneal shunt) with placement of a medium-pressure valve

was admitted for abnormal head movements in the form of "yes-yes" head movements. Clinical examination was normal.

Craniocerebral CT scan noted triventricular hydrocephalus and a cyst extending from the pineal region to the third ventricle, exerting a mass effect on the cerebellum (Figure 4).

We indicated endoscopic treatment, which consisted of a cysto-ventriculostomy.

The postoperative course was favorable, marked by an improvement of the abnormal movements from the first postoperative days. The control made 6 months later noted a complete amendment of the abnormal movements.



**Figure 4.** CT images of case N°3 **a):** axial section, **b):** sagittal reconstruction showing triventricular hydrocephalus and a cyst of the septum pelucidum extending from the pineal region to the third ventricle and compressing the cerebellum.

### DISCUSSION

Bobble head doll syndrome is a disorder of children under 10 years of age, rarely seen in adults. It is characterized by a stereotypy of back-and-forth movements of the head of 2-3Hz and inconstantly of laterality movements. In some cases the movements may extend to the shoulders, all upper limbs and trunk [9]. These movements are rhythmic, may intensify during stress, but are partially controllable by the patient and disappear during sleep [11]. At least 81 other cases have already been described before our cases [2,3,5,8,14,15,17,20] which have a particular clinical character due to the predominance of "non-non laterality movements". Since the first description in 1966[1], the consistency of intracerebral cystic abnormalities has been noticed but the direct causal link remains for the moment subject to controversies. The intracranial anomalies commonly observed are non-communicating hydrocephalus, supra sellar arachnoid cyst, septum

pellucidum cyst, choroid plexus papilloma, midbrain aqueduct stenosis and by extension all lesions of topography likely to lead to chronic obstructive hydrocephalus. These cystic lesions constitute valves that close the midbrain aqueduct[11,19,23].

The precise physiopathology remains unknown until now but many authors recognize the involvement of dysfunctions of the interconnections between the frontal lobes, the corpus callosum and the basal ganglia[16,17]. On this subject, several hypotheses have been formulated.

In the case of supra sellar cysts, some authors stipulate that the movements of the head would aim at tilting the cyst making the valve backwards in order to allow the opening of the mesencephalic aqueduct. Thus, the movements are interpreted as adaptation mechanisms serving to promote the restoration of normal cerebral spinal fluid (CSF) circulation in order to lower the intracranial pressure[19,23].

For other authors, the perpetual and rhythmic movements of the intracystic fluid or CSF on the diencephalon, the dorso-medial red nucleus and the motor pathways in the upper part of the midbrain would lead to dysfunctions of the extrapyramidal motor pathway[11,17,23]. The patient's ability to control movements and their disappearance during sleep would be arguments that support this hypothesis.

In our series we noted 2 cases of supra sellar cysts interfering with the circulation of the CSF; in addition we noted diffuse intracerebral calcifications which suggest sequelae of TORCH (Toxoplasmosis, Other Agents, Rubella, Cytomegalovirus, and Herpes Simplex) infection including cytomegalovirus infection[6,18,22] in the pediatric age. These same infections could also lead to meningitis and hydrocephalus as well as ocular manifestations including cataracts [12,18,21,22]. The symptomatology may be enriched by other signs related to intracranial hypertension.

The development of CT and MRI scans has improved the diagnostic time. Early diagnosis allows to avoid the permanent installation of the consequences of intracranial hypertension[9] such as blindness and endocrine disorders. Surgical management combines several techniques depending on the causal lesion: ventriculocisternostomy, cysto-ventriculoperitoneal shunt, cystectomy, endoscopic or open

marsupialization [10,11,13]. In our case the indicated techniques were ventriculocisternostomy, cysto-ventriculostomy and ventriculo-peritoneal shunt.

The postoperative results are generally marked by a significant clinical improvement[10,20]. On the other hand, there is still a residual risk of the usual complications linked to the type of operation and the approach. Recurrences are rare[20], estimated at 11%, and may be the consequence of obstruction of the ventriculocisternostomy orifices, dysfunction of the ventriculoperitoneal shunt system or postoperative ectasia of the fourth ventricle[11,4]. In one of the cases in our series (case N° 1) early clinical improvement was observed but shortened due to the probable closure of the ventriculocisternostomy orifices. The occurrence of the chronic subdural hematoma would result from a possible hyper draining following the second ventriculocisternostomy.

## CONCLUSION

Bobble head doll syndrome is a rare pediatric clinical entity, responsible for abnormal movements. It is a manifestation of the consequences of chronic hydrocephalus, which, if managed well and early, leads to favorable results. The quality of the clinical improvement depends on the diagnostic delay and the success of the etiological procedure. Our series of cases are typical examples that have been effectively managed with favorable results.

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