



# Bone Malformation of the Cervico-Occipital Causing a Basilar Impression and Triventricular Hydrocephalus

Tolojanahary Herizo Andrianjakamanana<sup>1\*</sup>, Ny Ony Narindra Lova Hasina Rajaonarison<sup>1</sup>, Hasina Dina Ranoharison<sup>2</sup>, Emmylou Prisca Gabrielle Andrianah<sup>1</sup>, Ahmad Ahmad<sup>1</sup>

<sup>1</sup>Departement of Radiology of the Joseph Ravoahangy Andrianavalona Hospital (CHUJRA), Antananarivo, Madagascar

<sup>2</sup>Departement of Radiology of the Andohatampenaka Hospital, Antananarivo, Madagascar

Email: \*tolojanahariherizo@gmail.com

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## Abstract

Congenital bone malformations of the cervico-occipital hinge are rare entities. They are often asymptomatic and often incidentally discovered during imaging examinations for malformative assessment or for other indications, but they can be the cause of neurological symptoms or signs of compression in the posterior fossa. We report a case of a bony malformation of the cervico-occipital hinge in a 16-year-old girl, with no medical and surgical history or known history of trauma. This malformation associates hypertrophy of the odontoid with basilar impression, causing triventricular hydrocephalus. The diagnosis was made by the cervico-cerebral scanner requested in front of a gait disorder and an attitude of cervical hyperextension since early childhood.

## Subject Areas

Pathology

## Keywords

Cervico-Occipital Hinge, Congenital Malformation, CT Scan, Hydrocephalus, Odontoid

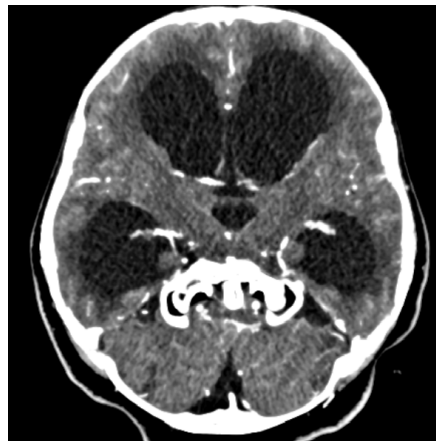
## 1. Introduction

The bone malformations of the cervico-occipital hinge are mainly represented by the basilar impression, defined by an odontoid process whose point reaches a level clearly higher than that of the line connecting the bony palate and the posterior edge of the foramen magnum (line of Chamberlain or Mac Gregor) on

a sagittal plane of a CT scan or conventional MRI [1]. These bone malformations are often associated with hyperplasia of the odontoid bone, also called dolichoodontoid. Bone abnormalities of the cervico-occipital hinge are rare, unlike acquired pathologies such as Paget's disease. The discovery is often fortuitous, because they are often asymptomatic, but there are also serious forms linked to neurological complications and compression syndromes in the posterior fossa as well as hydrocephalus [2]. We report a case of congenital bone malformation of the cervico-occipital hinge in a 16-year-old girl, characterized by odontoid hyperplasia with basilar impression, causing triventricular hydrocephalus diagnosed on brain CT. Our objective through this manuscript is to describe the clinical and CT appearance of this rare pathology, but potentially serious by its complications.

## 2. Observation

This is a 16-year-old girl, born vaginally in vertex presentation, without complication or instrumental extraction maneuver, with good neonatal adaptation, but presenting with cervical hyperextension from birth. She had no history of seizures or cerebro-meningeal infection. There was no history of traumatic brain injury or cervical trauma. His parents brought him in consultation for a gait disorder and an attitude of cervical hyperextension since early childhood. She also complained of a recurrent chronic headache. On physical examination, she was conscious, with a Glasgow score of 15/15, with a stable hemodynamic state, a fairly good general condition, afebrile, and the neurological examination had not objectified any sensory-motor deficit. The remainder of the physical examination was unremarkable. The cerebral scanner without and with injection of iodinated contrast product showed dilation of the lateral ventricular cavities and of the third ventricle, of homogeneous non-partitioned hypodense content, evoking tri-ventricular hydrocephalus (Figure 1), whose Evans score is calculated

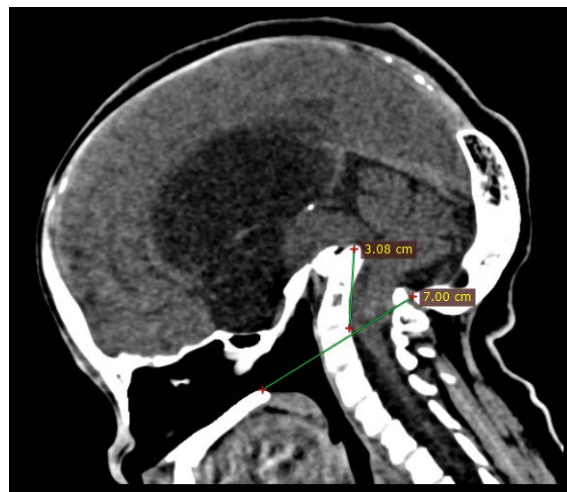


**Figure 1.** Axial section of a cerebral CT scan with injection of iodine product, in the arterial phase, showing dilation of the lateral ventricles and of the third ventricle.

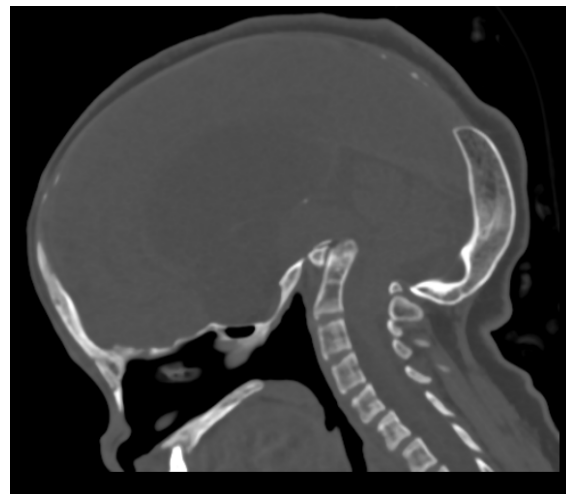
at 0.55, with signs of transependymal resorption. There was no visible obstruction at the foramen of Monro or posterior fossa mass. The odontoid is hyperplastic, protruding 30 mm above Chamberlain's line with a curvilinear aspect behind, pushing back the brainstem and compressing the fourth ventricle (**Figure 2** and **Figure 3**). A symptomatic surgical treatment by a ventricular bypass was undertaken with a partial improvement of the headaches.

### 3. Discussion

Congenital bone malformations of the cervico-occipital hinge are rare pathologies. Very few cases are described in the literature [3]. Basilar impression is characterized by upward translocation of the upper cervical spine and clivus into the foramen magnum, related to abnormal skeletal development. This is the most



(a)



(b)

**Figure 2.** sagittal reconstruction of a cerebral CT scan in the parenchymal window (a) and in the bone window (b), showing a protrusion of the odontoid, exceeding Chamberlain's line (green line).



**Figure 3.** Sagittal reconstruction of a brain scan, showing a hyperplastic odontoid, compressing the brainstem and the fourth ventricle.

common of these bone malformations [1]. They are often associated with hypertrophy of the odontoid or dolichodontoid, the etiopathogenesis of which is poorly understood, but congenital or traumatic etiologies are discussed [2]. These lesions are often asymptomatic, but can cause serious neurological complications. In our case, the clinical symptoms were present from early childhood but they were neglected, hence the delay in diagnosis, which was not established until the age of 16 years. The absence of a history of obstetrical trauma or crani-encephalic trauma in our patient suggests the congenital malformation origin of this pathology, given that there are no other causes of acquired anomaly of the hinge, cervico-occipital disease such as Paget's disease or gigantism [1]. The clinical manifestations were a vicious attitude of the neck, linked to the disorder of cervical statics and clinical manifestations related to hydrocephalus, but the patient did not present deficit neurological manifestations such as those described in the literature [2]. Bone malformations of the cervico-occipital hinge are often associated with other malformations such as Chiari disease or other anomalies such as neural tube closure anomalies. In our case, only bone malformations were the cause of the complications. Hydrocephalus is more common in other cervico-occipital hinge malformations than in isolated bone malformations [4]. Diagnosis is based on cross-sectional imaging techniques such as magnetic resonance imaging (MRI) [1] or CT scan [5], which also detects spinal cord and associated brainstem abnormalities. In our case, the cerebral scanner allows to establish the diagnosis, highlighting a tri-ventricular hydrocephalus, the origin of which is a compression of the fourth ventricle and the brainstem by the hypertrophic and protuberant odontoid, which is visible on the sagittal reconstruction. Basilar impressions or basilar invaginations are classified into two types depending on the presence (Type I) or absence (Type II) of Chiari disease [6]. The treatment of congenital malformations of the cervico-occipital hinge is mainly neurosurgical [7]. In our case, the treatment was above all symptomatic, by setting up ventricular drainage to alleviate the hydrocephalus.

## 4. Conclusion

Bone malformations of the cervico-occipital hinge are rare and often asymptomatic, diagnosed incidentally during CT scans or cranio-cervical MRI. They may also be the cause of clinical manifestations, due to compression of the structures of the posterior fossa or the origin of a hydrodynamic disorder of the cerebrospinal fluid, resulting in hydrocephalus, which is a serious complication. In front of a scanographic or MRI assessment of a malformation of the cervico-occipital hinge or in front of an etiological search for hydrocephalus, it is imperative to look for a bone malformation of the cervico-occipital hinge, by performing a sagittal reconstruction.

## Conflicts of Interest

The authors declare no conflicts of interest.

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