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*Research Article*

# Congenital Malformation of the Posterior Cerebral Pit Type Dandy-Walker: About A Case

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## Introduction

Dandy-Walker syndrome is a rare congenital malformation of the posterior cerebral fossa of unknown etiology. The first autopsy description was reported in 1887 by Sutton [1] and it was not until 1914 when Dandy and Blackfan [2] studied an association between hydrocephalus and cystic dilatation of the fourth ventricular. The malformation was further characterized by Dandy in [3] 1921 and by Taggart and Walker in [4] 1942 as being related to congenital atresia of the foramen of the fourth ventricle.but several studies indicate that there is a causal relationship with various types of chromosomal abnormalities and malformation syndrome. Its incidence is of the order of 1 case per 100,000 live births and it is more common in boys (1.24 per 100,000) than in girls (0.78 per 100,000).

## Methodology

This was a descriptive cross-sectional study about a case of Dandy-Walker syndrome diagnosed in the neurology department of the Sino-Central African Friendship University Hospital in Bangui.

## Medical Observation

This was the patient G.D, aged 21, student, hospitalized on November 20, 2021 in the neurology department of the Sino-Central African Friendship University Hospital for: headaches, motor deficit of the left hemibody, disorder of vigilance and vesico-sphincter type of urinary incontinence and anal retention. The start of the symptoms dates back to the morning of November 2, 2021 with the rapidly progressive onset of headaches of progressively increasing intensity. In the evening, the headaches became boring and insomnia. Faced with these signs, self-medication was carried out with paracetamol tablets 1g every eight hours, leading to partial remission of the headaches after five days. On the morning of November 20, 2021, upon waking up, the patient presented with left hemibody heaviness, urinary incontinence, anal retention followed during the day by impaired vigilance, thus motivating an emergency consultation in neurology for better investigation and management charge. His pathological history was unremarkable. The neurological examination showed: obtundation with Glasgow score of 13/15, BP was 140/100 mmHg, rotatory nystagmus in extreme gaze on both sides, divergent strabismus of the left eye, left hemiparesis not proportional with FM = 0/5 at the MS and 2/5 at the MI, the bicipital and patellar ROT are abolished on the ipsilateral hemibody, the Babinski sign is indifferent on the left. The remainder of the examination was unremarkable.

The para-clinical examination showed: leukocytosis at 14100/mm$^3$,

ESR at 130 mm at the 1st hour, SLV negative, BW negative, hepatic cytolysis with AST at 215 IU/l and ALT at 404 IU/l, Gamma GT at 91 IU/l, Klebsiella oxytoca cystitis;

- Brain CT was in favor of a supratentorial arteriovenous malformation with associated bleeding; at the infratentorial level: a vermian cerebellar malformation and a cystic one leading to tentorial elevation.
- The clinical course was marked by the occurrence of anemia requiring a transfusion of two bags of 450 ml of whole blood iso rhesus group on 11/20/2021 and a death occurring on 11/25/2021 in relation with intracerebral hemorrhage due to probable arteriovenous rupture, one day after the result of the brain CT was available, allowing the postmortem diagnosis to be established.

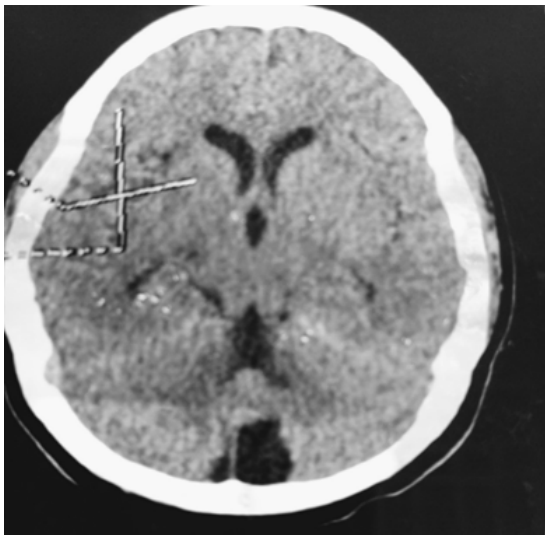

**Image 1:** Source National Center for Medical Imaging 2021 (Central African Republic).

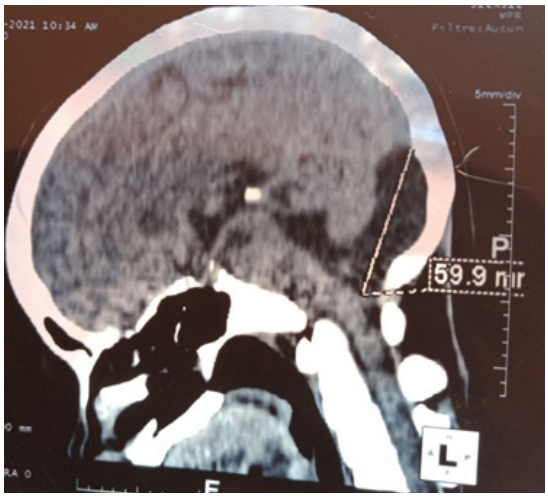


**Image 2:** Source National Center for Medical Imaging 2021 (Central African Republic).

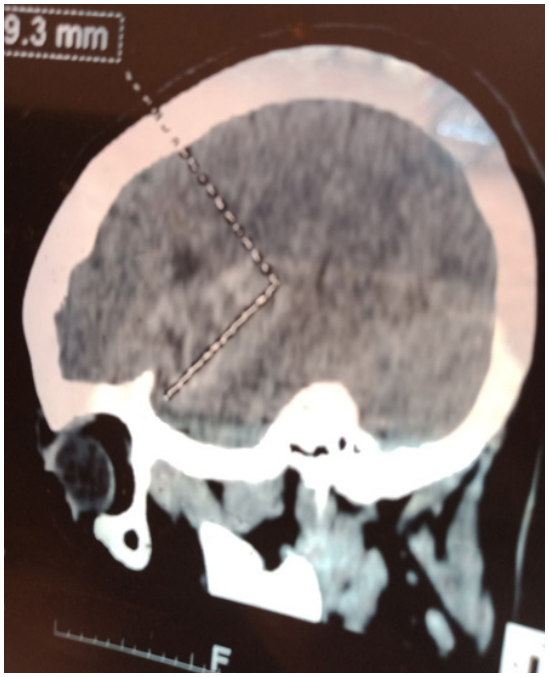


**Image 3:** Source National Center for Medical Imaging 2021 (Central African Republic).

## Discussion

Dandy-Walker syndrome is a malformation of the central nervous system. Sutton made the first description of the malformation in 1887 [5], describing hydrocephalus associated with a posterior fossa cyst and hypoplasia of the vermis. It was in 1954 that Benda [6] first introduced the term Dandy Walker syndrome. He defines it as a syndrome with cerebellar involvement with dysgenesis of the vermis, an enlargement of the posterior fossa containing a cystic formation with an enlargement of the IVth ventricle and a raised position of the tentorium of the cerebellum.

Peripheral malformations associated with Dandy-Walker syndrome are often described, the frequency of which varies greatly and can reach 76% [7]. In our case, such peripheral damage was also present in the form of divergent strabismus of the left eye, which affects 10.5% of Dandy-Walker syndrome carriers [8]. In adults, progressive and slow decompensation can manifest itself as: headaches, especially chronic headaches, balance difficulties, walking disorders, unilateral sensory disorders, syncope, a progressive decrease in cognitive and intellectual abilities suggesting a picture of dementia, multiple psychiatric disorders [9].

In our case, the patient had symptomatic Dandy-Walker syndrome.

- The headaches were certainly related to intracranial hypertension;
- The vigilance disorder could be linked to acute decompensation due to probable damage to the brainstem;

- Nystagmus could reflect either a visual disorder, therefore in relation to divergent strabismus of the left eye or in relation to cerebellar damage, therefore in direct relation to Dandy-Walker syndrome;
- Non-proportional left hemiparesis, which could be explained by damage to the subcortical motor pathways.

The reasons for this asymptomatic period, as well as the reasons for progressive decompensation of the syndrome, remain uncertain [9]. For some authors, the reasons seem to lie in the existing imbalance between the production of cerebrospinal fluid and its absorption, which occurs late in adults [10].

Acute decompensation is manifested by: nausea, vomiting, headache, nystagmus, papilledema, etc. It is often linked to a sudden increase in intracranial pressure [11]. At the central nervous system level, DW syndrome is associated with other malformations in 48% [12] of cases respectively. In our case, we note as central nervous system malformations, a supratentorial arteriovenous malformation with bleeding and intracranial hypertension.

## Conclusion

Dandy-Walker syndrome is part of the group of posterior fossa malformations. It can be latent or symptomatic which can lead to serious neuropsychological manifestations in young subjects. In children, it can cause a delay in psychomotor acquisition. The treatment is surgical and consists of drainage of the cyst and/or hydrocephalus. This observation should draw the attention of health providers to the importance of brain CT in the face of neurological deficit, signs of intracranial hypertension and/or hydrocephalus in young subjects and children.

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