# The Menace of Neglected Hydrocephalus – Case Report And Review Of Literature

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**Abstract**

**Introduction**
Timely diagnosis and management of hydrocephalus are crucial to mitigating adverse outcomes. The etiology of hydrocephalus in infants is diverse, while the clinical presentation of hydrocephalus in infants can be varied. Neuroimaging is crucial in diagnosis. Neglected hydrocephalus poses an even more serious challenge. There is a high risk of scalp erosion and wound dehiscence following a ventriculoperitoneal shunting in very a stretched scalp. We present a case recently managed in our centre to highlight the menace of neglect.

**Case Summary/Result**
A 7-month-old baby presented with a history of progressive enlargement of the head for five months duration. There was also poor progress in developmental milestone attainment. Antenatal history was unremarkable. Occipitofrontal circumference was 75cm (the normal range for 7 months was 42.5 -46.5cm). He was worked up for ventriculoperitoneal shunting, which he eventually had but developed shunt erosion and the overriding skull bones postoperatively

**Conclusion**
Neglected hydrocephalus is still a problem in our environment. Late presentation leads to unwanted complications.

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## Introduction

Timely diagnosis and management of hydrocephalus are crucial to mitigating adverse outcomes. Hydrocephalus, defined as the abnormal accumulation of cerebrospinal fluid (CSF) within the brain's ventricular system, poses a significant threat to the neurological development of children, especially infants.[1,2] It can lead to increased intracranial pressure, progressive ventricular dilation, and subsequent brain damage if left unattended.[1] It is estimated that the incidence of congenital and acquired hydrocephalus is between 80 and 125 per 100,000 births depending on the regions.[3] In our clime, this constitutes a significant burden in paediatric neurosurgery.[4,5]

The etiology of hydrocephalus in infants is diverse, encompassing congenital malformations (e.g., aqueduct stenosis), intraventricular hemorrhage, infections (e.g., meningitis), and neoplasms.[6,7] Each of these etiologies presents distinct diagnostic and therapeutic challenges.

The clinical presentation of hydrocephalus in infants can be varied and often includes nonspecific symptoms such as irritability, poor feeding, vomiting, lethargy, and an increasing head circumference. Since these symptoms can be subtle and easily misinterpreted, there could be a delay in recognition which can result in significant neurological compromise, highlighting the need for early intervention and robust healthcare infrastructure.

Neuroimaging, primarily through cranial ultrasound in neonates and magnetic resonance imaging (MRI) or computed

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tomography (CT) scans in older infants, remains the mainstay of diagnosis, providing essential information on ventricular size and possible causes of obstruction.[6,8,9,10]

Management of hydrocephalus typically involves surgical intervention to divert CSF and relieve intracranial pressure. In our clime and most African countries, the most popular methods include ventriculoperitoneal (VP) shunt placement and endoscopic third ventriculostomy (ETV).[11,12] Each technique has its indications, benefits, and risks. VP shunts, while commonly used, are associated with complications such as infections, obstructions, and the possible need for multiple revisions over a patient's lifetime. ETV, although less invasive, is not suitable for all types of hydrocephalus and carries its own set of risks, including the potential for inadequate CSF diversion. [13]

Neglected hydrocephalus poses an even more serious challenge. Unfortunately, it is still a problem in low-income countries. [14,15] These management challenges are compounded by the potential for advanced brain damage, increased intracranial pressure, and significant developmental delays.[16,17] The timing of intervention now becomes critical, though the window for optimal neurodevelopmental outcomes may have already been compromised. Additionally, the presence of severe neurological deficits at presentation can influence surgical decisions as well as postoperative care strategies.[18,19]

Besides the risk of external rupture reported by some authors, extreme cortical thinning may limit the neurological outcomes. Besides, there is a higher risk of hardware erosion and wound dehiscence as a result of an extremely stretched scalp. Other complications like peri-catheter leakage, subdural hematoma, over-riding skull bones and shunt migration are known complications.[20]

We present a case of neglected hydrocephalus recently managed in the unit in order to raise the awareness for this unfortunate condition.

**Case Summary**

A 7-month-old baby presented with a history of progressive enlargement of the head for five months duration. This was associated with the downward rolling of the eye and intermittent vomiting. There was also poor progress in developmental milestone attainment as the baby was yet to achieve head control at presentation.

He was said to have developed recurrent fever, vomiting, and recurrent focal tonic seizure involving the right upper and lower limb at 2 months of age for which the mother presented to a peripheral hospital. The patient was said to have been treated but subsequently had progressive head enlargement after discharge. The parents resorted to herbal medications which were applied to the head. This continued until the patient was then presented on account of massive head enlargement and inability to control the head.

Pregnancy was spontaneously achieved, registered for Antenatal care at 16 weeks gestational age, and took medications as prescribed. No history of fever or use of over-the-counter drugs. Pregnancy was carried to term and delivered via spontaneous vaginal delivery. The patient's weight at birth was 2.3kg. There was no perinatal fever, jaundice, or untoward event.

On examination, the vital signs were normal. Baby was active Occipitofrontal circumference was 75cm (the normal range for 7 months was 42.5 -46.5cm). See figure 1

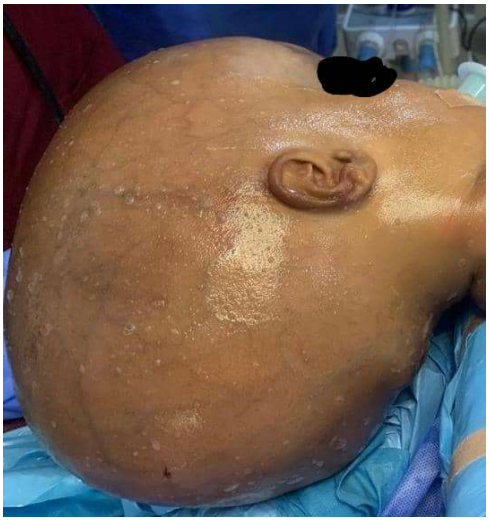

Figure 1: showing head enlargement.

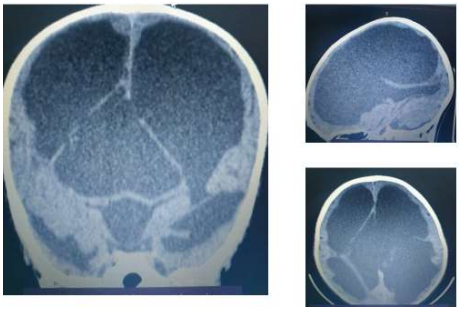

Fig 2: Showing CT scan images of massive hydrocephalus with triventriculomegaly.

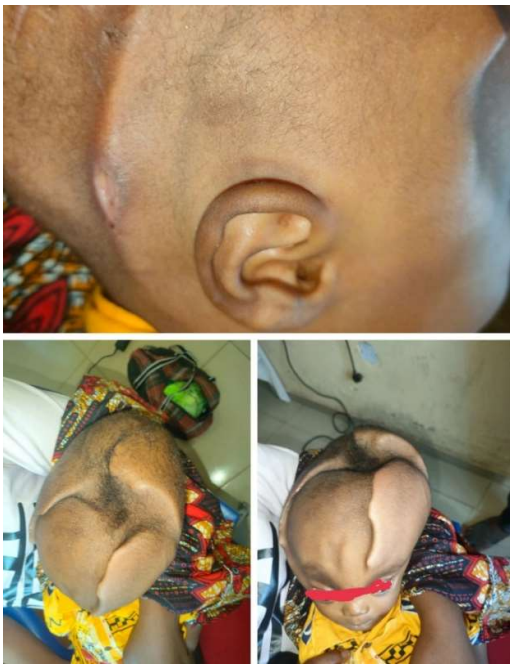

Figure 3: showing shunt erosion and the overriding skull bone

Patient had a cranial CT (see figure 20), septic and other blood workup. Patient eventually had a ventriculoperitoneal shunting.

**Discussion**

Hydrocephalus is a common paediatric neurosurgical condition with significant neurological sequelae. It is a leading cause of disability among children worldwide. [21]

CSF diversion commonly done in our clime includes Endoscopic Third Ventriculostomy (ETV) and Ventriculoperitoneal shunting (VPS).

Endoscopic Third Ventriculostomy (ETV) and its adjuncts as a management option have the benefit of obviating the need for an implant that would be carried for life. Hence, it potentially has lower complication rates. [21] Unfortunately, there is limited availability of these modalities in most neurosurgical centres in Nigeria. [22]

Ventriculoperitoneal shunting is widely used in Nigeria in centres where ETV is not available or where the patients fail to meet the criteria for ETV. [23,24] However, VPS is associated with several complications which may occur from the time of surgery through several years later. These complications include the following [25]:

- Infection due to skin flora entering the shunt, usually from *Staphylococcus epidermidis*
- Intracerebral or intraventricular haemorrhage
- Malposition
- Abdominal perforation during placement
- Shunt erosion of the skin with exposure of the system
- Shunt over drainage (slit ventricles)
- Shunt nephritis
- Shunt disconnection
- Shunt obstruction
- Subdural haematomas
- Abdominal CSF collections (pseudocyst or CSFoma)
- Shunt breakage at any point

- Catheter perforation of viscera and rarely extrusion through the anus
- Inguinal hernia
- Seizures.

The neglect of patients with hydrocephalus may be associated with the arrest of hydrocephalus or continued head enlargement. [26] Neglected maximum hydrocephalus as seen in our index patient is usually associated with severe stretching of the scalp resulting in an increased risk of scalp necrosis and shunt exposure. (see figure 3). The risk of infection is also increased. There is also the problem of unsatisfactory cosmetic results after CSF diversion due to the rapid decompression of the head and awkward alignment and the overriding of the skull bones. (see figure 2). Besides these complications, the issue of optimum position for surgery and airway management intraoperatively also poses a significant challenge.

Several reasons have been alluded to as responsible for late presentations of patients with hydrocephalus. These have ranged from poverty, ignorance, religious/local belief, delayed referral to a centre with Neurosurgical facilities and previous ugly experiences with patient care in hospitals. [27] Subsidizing the management of hydrocephalus, massive enlightenment campaigns and improving the overall quality of healthcare services in our healthcare system are proven methods of encouraging early presentation and improving outcomes of the management of hydrocephalus patients.

**Conclusion**

Neglected hydrocephalus is still a problem in our environment. Late presentation leads to unwanted complications. There is a need for increased awareness of the benefit of early presentation.

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