












Acute hydrocephalus following acute polyradiculoneuritis in pediatric patients: observation of 2 cases treated with ventriculoperitoneal shunt and systematic review of the literature

Authors: Hugues Ghislain Atakla^{1,2,5} , Mohameth Faye^{1,2} , Aichat Yabo Salami^{3,5} , Patrick Ewusi Bankah⁴ , Bob Makuta^{1,2} , Reginald Nsiah⁴ , Assia Kahkahni¹ , Irene Wrokie Kweidjartey⁴ , Dismand Stephan Houinato⁵ , Alioune Badara Thiam¹ , Momar Code Ba¹ 

Affiliations

1. Neurosurgery Department, Fann's national teaching hospital center, Dakar, Senegal
2. Neurosurgery Department, Idrissa Pouye General Hospital, Dakar, Senegal
3. Biological medical laboratory, national teaching hospital centre, Cotonou, Benin
4. Neurosurgery Department, Korle-bu teaching hospital, Accra, Ghana
5. Laboratory of Noncommunicable and Neurologic Diseases Epidemiology, Faculty of Health Science, University of Abomey-Calavi, Cotonou, Benin

Corresponding Author: Hugues Ghislain Atakla. Email: neuroscience.ghislain@gmail.com

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Abstract

Background: Acute polyradiculoneuritis, or Guillain-Barré syndrome, is an immune-mediated disorder of the peripheral nerves that typically presents with ascending weakness. Central complications are rare in children, with acute communicating hydrocephalus being exceptionally uncommon. This form of hydrocephalus, caused by impaired CSF resorption despite preserved flow, can be fatal if untreated. To our knowledge, this is the first reported pediatric case of such a complication in Africa. **Case report:** We report two cases of children who were managed in the neurosurgery department for acute communicating hydrocephalus. Both had been treated a few days earlier in the pediatric neurology unit for acute polyradiculoneuritis. Each patient underwent ventriculoperitoneal (VP) shunt placement combined with medical therapy, resulting in favorable clinical outcomes. **Conclusion:** Acute hydrocephalus is a rare but serious complication of Guillain-Barré syndrome in children. It may be exacerbated by velopharyngeal paralysis, respiratory failure, and the limited ability of young children to articulate their symptoms. Early diagnosis and prompt surgical intervention, particularly VP shunting, are critical and potentially life-saving.

Key words: Hydrocephalus, acute polyradiculoneuritis, child, ventriculoperitoneal shunt.

BACKGROUND

Acute polyradiculoneuritis (acute PRN), also known as Guillain-Barré syndrome (GBS), is an immune-mediated disease of the peripheral nerves that has been clinically well known since 1916. It can occur at any age and its main feature is acute, rapidly ascending flaccid paresis [1]. Peak paralysis usually occurs two weeks after the onset of the disease. There is often a history of infection in the weeks preceding the onset of motor symptoms. [1] Children usually recover from the motor dysfunction however, autonomic instability and respiratory impairment may occur requiring close monitoring and even admission to an intensive care unit. [2]

Typical complications of the central nervous system (CNS) are rarely reported in the literature. Hydrocephalus (HCP) is a progressive distension of the anatomical spaces (ventricular and subarachnoid) where the cerebrospinal fluid (CSF) normally resides. [3] When it is not due to excessive production or an obstruction to the circulation

of CSF, it is probably linked to a defect in the resorption of CSF. [4] Taking into account the data currently available in the literature, the link between HCP and acute PRN is not clear.

According to some authors, hydrocephalus and increased intracranial pressure are uncommon but well-known complications of acute PRN, occurring in approximately 4% of cases [5,6]. The widely accepted underlying mechanism for HCP formation is reduced CSF absorption due to elevated protein concentration blocking the arachnoid villi [7].

Based on a systematic review of the literature on active hydrocephalus and acute PRN in the paediatric population, we noted that no cases have yet been reported in Africa. These are therefore the first 2 cases we are reporting in the African context. In this study, we describe two (2) cases of active hydrocephalus in the setting of acute PRN in subjects of paediatric age for whom we performed ventriculoperitoneal shunt (VPS) surgery with a good clinical outcome.

METHODS

Data selection and search strategy

A systematic review was carried out. PubMed and Google Scholar were searched. All available literature on acute HCP in the context of acute PRN in paediatric patients and published before 30/10/2024 was indexed. Two authors, H.G. Atakla and M. Faye, independently selected abstracts and full texts, resolving the following questions: countries with reported studies of acute communicating HCP secondary to acute PRN, frequency of acute communicating HCP secondary to acute PRN in these countries, overall management, and overall prognosis.

Inclusion criteria and study selection

Studies were included if they reported results related to acute non-obstructive HCP and subsequent to acute PRN. We included

subjects of all ages in the systematic review with close attention to studies involving pediatric-aged subjects. No restrictions were placed on study design or language of publication. Studies for which the full text was not available were excluded.

Clinical and radiological aspects

There are a number of internationally recognised schemes and guidelines for the diagnosis of acute PRN and acute HCP. A careful history was sufficient to suggest acute PRN, and the result of CSF analysis showed typical albuminocytological dissociation (high protein level with normal cell count) with a characteristic electroneuromyogram. Acute communicating HCP, on the other hand, manifests itself on cerebral CT scan as complete dilatation of the various ventricular

systems, with no visible impediment to CSF circulation. When this dilatation is significant and active, signs of trans-ependymal resorption can be observed at the periventricular level.

Data extraction

Study and patient characteristics, including first author, year and country of publication, study type, patient age and percentage of acute communicating HCP following acute PRN, were collected. All data were collected independently by the two reviewers and discussed scientifically, free of any conflicts of interest.

CLINICAL CASES

Case Number 1

A 9-year-old boy, with a past history of SARS-CoV-2 infection two years prior and an up-to-date vaccination status, was being followed in the pediatric neurology department for acute Guillain-Barré syndrome (GBS)-type polyradiculoneuritis (PRN), for which he was receiving intravenous methylprednisolone. Twenty-six days after the onset of symptoms, he developed a progressively worsening, persistent headache associated with vomiting, excessive sweating, respiratory distress, and altered consciousness. These developments prompted an urgent referral to the neurosurgery department. According to the history provided by his parents, there had been no recent vaccination, injection, or documented infectious illness.

Clinical examination revealed features consistent with acute intracranial hypertension (ICH), including headache, vomiting, diplopia, and sixth cranial nerve (CN VI) palsy. Neurological assessment showed a peripheral neurogenic syndrome with generalized motor weakness (Medical Research Council grade 3/5 in all four limbs), diffuse hyperesthesia, dysautonomia, facial diplegia, and dysphagia.

A brain computed tomography (CT) scan demonstrated active communicating

Statistical analysis

The various data were first presented descriptively. We performed a meta-analysis in R software (version 4.1.0) (Core Team, 2020) using the meta package (Balduzzi et al., 2019). Data were pooled with a random-effects model using the DerSimonian-Laird estimator for variance between studies. Publication bias was assessed using Egger's and Begg's tests, and funnel plots were drawn. No meta-regression was performed.

hydrocephalus, with dilation of the entire ventricular system and enhancement of cortical sulci and the skull base following contrast administration (Figure 1a–d). Fundoscopic examination revealed bilateral papilledema. A nasopharyngeal swab for SARS-CoV-2 by RT-PCR was negative.

Given the clinical and radiological findings, an emergency external ventricular shunt (EVS) was placed, and cerebrospinal fluid (CSF) was obtained for cytochemical, bacteriological, and virological analyses. The preoperative CSF appeared clear, with an opening pressure of 60 cmH₂O. CSF analysis revealed albuminocytologic dissociation characterized by elevated protein concentration (64 mg/dL) with normal glucose and lymphocyte counts. No pathogens were identified.

Within 48 hours, the patient regained consciousness and spontaneous respiration, allowing for extubation. Due to the infection risk associated with prolonged external drainage, an early conversion to a ventriculoperitoneal shunt was performed..

The postoperative course was uneventful, with complete resolution of intracranial hypertension. Parenteral treatment included

analgesics, methylprednisolone at 40 mg/day, and adjuvant therapies. Six weeks after symptom onset, the patient exhibited motor recovery graded at 4/5 in all four limbs, along with persistent sensory disturbances and signs of autonomic dysfunction.

At the 12-month follow-up, the patient demonstrated full recovery across all functional domains. There were no residual symptoms such as fatigue, gait abnormalities, or sensory deficits.

Case Number 2

A 5-year-old girl was admitted to the emergency department with progressive weakness in all four limbs, diffuse hypersensitivity, and marked irritability. History obtained from the parents indicated that the patient's vaccination schedule was up to date, with no recent vaccinations, injections, or identifiable infectious illness.

She was admitted to the pediatric ward, where a neurology consultation concluded that the Guillain-Barré syndrome (GBS) had been evolving for approximately 13 days at the time of admission. Electroneuromyography (ENMG) revealed segmental demyelination without axonal involvement, decreased motor conduction velocities, prolonged distal latencies, and delayed F- and H-wave responses.

After six days of hospitalization (day 19 of symptom evolution), the patient developed acute headaches, projectile vomiting, and rapid clinical deterioration. On examination, her Glasgow Coma Scale (GCS) score was 12/15 (E3V4M5), and she exhibited intermediate, isochoic pupils with sluggish light reactivity.

An emergency brain computed tomography (CT) scan revealed active communicating hydrocephalus (Figure 2a–b), prompting immediate external ventricular shunting (EVS). Intraoperatively, the cerebrospinal fluid (CSF) appeared clear. CSF analysis showed mild protein elevation (proteinorachia: 0.5 g/L), normal glucose (glucorachia: 0.58 g/L), and a leukocyte count of 8 cells/mm³—findings that were considered non-specific.

Subsequently, the external valve was internalized through placement of a ventriculoperitoneal shunt (VPS). The postoperative course was uncomplicated, with resolution of signs of intracranial hypertension. Neurological examination demonstrated progressive sensorimotor recovery over the following eight weeks. The patient was discharged in good general condition after six weeks of hospitalization. At six-month follow-up, she exhibited complete clinical remission with no residual neurological deficits.

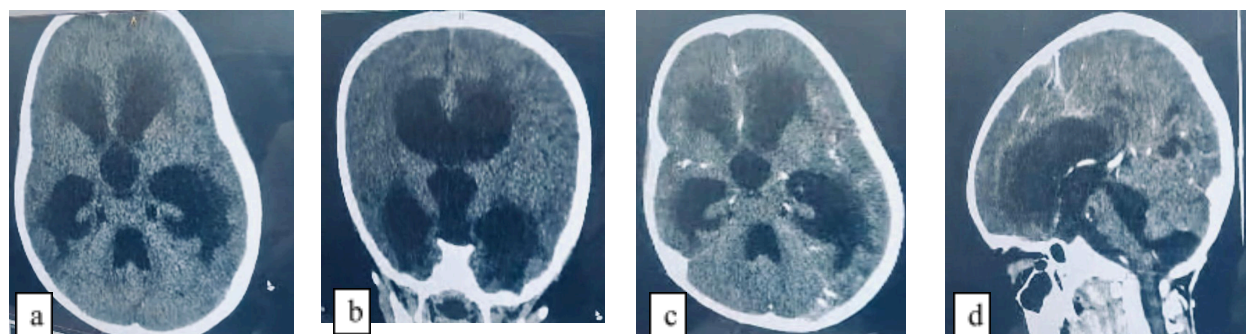


Figure 1: Contrast-free brain CT, axial section (1a) and coronal reconstruction (1b) showing active hydrocephalus with dilatation of all ventricular systems of the brain. Contrast enhanced axial slice (1c) and sagittal reconstruction (1d) showing raised cortical sulci and skull base, indicative of inflammation.

Table 1: Case summaries

Age/Gender	Medical history	Clinical Signs	Cerebral CT scan	Treatment	Outcome at 1 year post-op
Case 1 9 / M	Covid-19	ICH; Consciousness disorders; GBS	Acute communicating HCP	EVS first and VPS secondary Immunosuppressive therapies	Full recovery Normal neurological check-up
Case 2 5 years / M	None	ICH Consciousness disorders GBS	Acute communicating HCP	EVS first and VPS secondary Immunosuppressive therapies	Normal neurological check-up

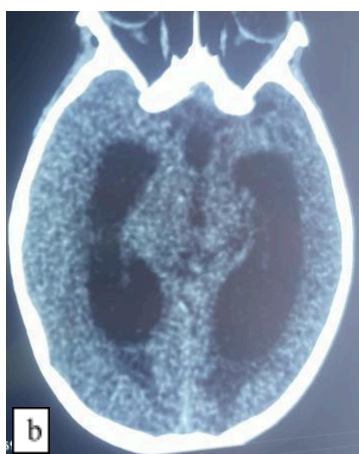
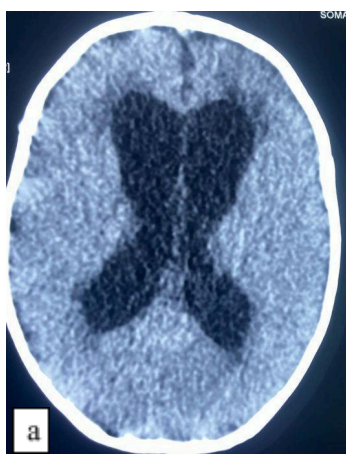


Figure 2a and b: Brain CT scan, parenchymal axial: active hydrocephalus with significant dilation of the temporal horns.

DISCUSSION

We presented two pediatric patients who were transferred to the neurosurgery department at different times for the management of active non-resorptive (communicating) hydrocephalus, occurring in the context of acute polyradiculoneuritis (PRN). This association is recognized as a rare complication of Guillain-Barré syndrome (GBS).

Autoantibodies directed against nerve gangliosides are thought to play a central role in the pathogenesis of many GBS cases

[8]. Acute PRN is particularly uncommon in very young children, in whom the condition is often underrecognised due to its lower incidence, challenges in obtaining a reliable history, and difficulties in performing neurological examinations and electrophysiological studies, such as electromyography (EMG), which require specialized expertise in this age group [9,10].

Hydrocephalus is an exceptionally rare complication of PRN-type GBS and may occur at any stage during the disease

course. To our knowledge, beyond the two cases presented in this report, only thirteen additional pediatric cases have been documented over the past sixty years. The first reported case was published in Archives of Neurology in 1966 by Richard Janeway and colleagues [4], describing a child diagnosed and successfully treated in 1964. Since then, all reported cases in the literature have been isolated case reports, underscoring the rarity and clinical significance of this complication.

Several cases of hydrocephalus (HCP) following acute polyradiculoneuritis (PRN) have been reported in the international literature and are summarized in Table 3. Based on currently available data, the mean age of patients with GBS-associated hydrocephalus across all age groups is 17.92 years, ranging from 1 month to 68 years. The male-to-female ratio was 8:5. Of the 13 reported cases, 61.5% (8 patients) were of pediatric age, with an equal distribution between males and females (4 boys and 4 girls).

In addition to the symptoms of acute PRN, all patients—particularly children—exhibited signs of increased intracranial pressure (ICP). Hydrocephalus is a rare and potentially overlooked complication of acute PRN, especially in young children. The presence of respiratory difficulties and velopharyngeal paralysis, which often results in loss of verbal communication, may delay diagnosis and exacerbate the clinical course. For this reason, several authors recommend performing brain imaging in patients with GBS who present with respiratory failure, papilledema, or other signs suggestive of central nervous system involvement [12,22].

Three primary mechanisms have been proposed to explain the development of hydrocephalus in the context of acute PRN: (1) elevated cerebrospinal fluid (CSF) protein concentration and osmotic pressure, (2) impaired CSF resorption, and (3) cerebral edema [7,13,18]. Multiple studies, including those by Hantson P. [14], Reid A.C. [16], and

Farrell K. et al. [23], support the notion that CSF accumulation in the ventricular system is primarily due to defective resorption. This dysfunction is considered the principal mechanism leading to increased intracranial pressure and subsequent hydrocephalus.

In our cases, respiratory failure emerged as the most consistent predictor of underlying hydrocephalus, aligning with previous findings [12]. However, in both patients described in this report, signs of intracranial hypertension were the primary indication for neuroimaging, which ultimately confirmed the presence of hydrocephalus. Variability in clinical presentation, often seen in acute PRN, limits the diagnostic value of comparing specific signs across cases.

Due to the rarity of this complication and the lack of comprehensive, up-to-date literature, no standardized treatment guidelines exist for hydrocephalus secondary to acute PRN. Nevertheless, procedures such as the CSF puncture test or external lumbar drainage are recommended to help predict the success of shunt surgery [24]. It should be emphasized, however, that lumbar puncture has limited diagnostic value and may unnecessarily delay treatment when clinical and radiological findings are consistent. Once hydrocephalus is confirmed, prompt initiation of treatment—particularly surgical intervention—is essential to improving patient outcomes.

Although hydrocephalus associated with acute polyradiculoneuritis (PRN) may resolve spontaneously, the overall prognosis is generally favorable. In many cases, it can be managed successfully with medical therapy alone, and surgical intervention such as ventriculoperitoneal shunting (VPS) is rarely required [12,14,15,23]. Recovery time has been reported to range from 1 to 12 months in patients who underwent surgical intervention, and from 5 to 6 months in those treated conservatively with medical therapy [12].

In our cases, the severity of clinical symptoms at admission - despite initial

medical management - prompted us to proceed with systematic VPS. This intervention proved highly beneficial. The rapid resolution of signs of intracranial hypertension following shunt placement

confirmed the effectiveness of the surgical approach. Our findings are consistent with previously published data supporting early intervention in select cases [6,11,13].

Table 3: Cases of acute PRN with concomitant hydrocephalus reported in the literature.

	Authors	Patient age/Sex	Associated pathologies	References	Country
1	Abdulaziz Ata	23 years / F	None / NR	5	China
2	Erşahin Y	10 years /M	None / NR	6	Hungary
3	Richard Janeway	21 years / M	None / NR	11	USA
4	Liu Cy	68 years/M	Vaccination influenza	12	Taiwan
5	Gilmartin Rc	1 month / F	None / NR	13	USA
6	Hantson P	16 years / M	None / NR	14	Belgium
7	Barzegar M	21 months / F	None / NR	15	Iran
8	Reid Ac	16 years / M	None / NR	16	England
9	Ozdemir O	32 years / M	None / NR	17	Turkey
10	Morley Jb	46 years / M	None / NR	18	USA
11	Christina Doxaki	12 years /F	Obesity	19	Greece
12	Daniela Avila-Smirnow et al	9 months / M	Wiskott-Aldrich Syndrome	20	Chile
13	Mantadakis, Elpis MD	16 months / F	Infection with Bartonella quintana	21	Greece

NR: not reported; M: male; F: female

It is therefore reasonable to conclude that in the case of acute signs of ICH confirmed by active hydrocephalus on imaging in patients with acute PRN, internal shunt surgery is better and more beneficial for children.

The main limitations of this study were: The small size of our sample, and the scarcity or lack of recent scientific data on the topic. The absence of large-scale studies in paediatric settings.

Note and recommendations: Given the rarity and lack of awareness among many practitioners of this complication (acute HCP) in acute PRN, we remind clinicians in particular those in medical neurology and

pediatrics to pay close attention to this pathology, systematically looking for signs of ICH in the history especially in patients of pediatric age. Because there is a strong possibility that many cases occur in both children and adults that go undiagnosed for various reasons, leading to poor outcomes. Early diagnosis and prompt, appropriate management remarkably change the prognosis of this complication, which remains fatal without treatment. We believe that awareness-raising campaigns and transmission of scientific data within the scientific community are necessary. A subsequent study will envisage a global survey within several national health centres to identify cases and limit errant diagnoses.

CONCLUSION

Hydrocephalus is a rare but well-recognized complication of acute PRN. It can occur at any time during the disease progression and deserves special attention for early diagnosis and management. We present two cases of acute active hydrocephalus following acute PRN in two children who underwent emergency EVS and secondary VPS with a good clinical outcome. The clinico-radiological concordance was a major argument for shunt surgery. The unpredictable evolution of acute PRN and signs of ICH confirmed by active and severe HCP in a patient unable to express his pain motivate the performance of an internal shunt, which can prove highly beneficial, as in the 2 cases in this study.

Abbreviations:

CNS: central nervous system
CSF: cerebrospinal fluid
CT: computed tomography,
EVS: external ventricular shunt
VPS: ventriculoperitoneal shunt
GBS: Guillain-Barré syndrome,
IVIg : intravenous immunoglobulin,
LP: lumbar puncture.
ENG: electroneuromyogram
HCP: hydrocephalus
ICH: intracranial hypertension
MRC: medical research council
PRN: polyradiculoneuritis
NS: nervous system
VPS: Ventriculoperitoneal shunt

Contributions: All authors contributed to the study conception and design. Data collection

and analysis were performed by Hugues Ghislain ATAKLA, the first draft of the manuscript was written by Hugues Ghislain ATAKLA, and all authors commented on previous versions of the manuscript. All authors read and approved the final manuscript.

Ethics approval: The study was approved by the institutional review board of National teaching hospital Fann of Dakar. The patients or their relatives, provided informed consent for the inclusion of their clinical data in this study.

Consent to participate: Informed consent was obtained from the patient.

Consent for publication: Informed consent was obtained from the patient.

Conflict of Interest: The authors declare that the research was conducted in the absence of any commercial or financial relationships that could be construed as a potential conflict of interest. No competing interests.

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