

# A Report on Two Cases of Spontaneous Cutaneous Rupture Secondary to Severe Congenital Hydrocephalus

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## Abstract

**Introduction:** Congenital hydrocephalus is one of the most common causes of neurosurgical consults globally. Management delays may cause unique complications such as spontaneous cutaneous rupture and cerebrospinal fluid leakage. This may occur frequently in low-to-middle income countries, where prenatal and neurosurgical healthcare services are less accessible; however, no such cases have been recorded in the Philippines.

**Case Presentation:** We present two cases of severe hydrocephalus with spontaneous cutaneous rupture. The first is a 7 - month-old female with leaking CSF from a 1-centimeter spontaneous cutaneous rupture at the right anterior aspect of her anterior fontanelle. On admission, her head circumference was 85 cm. Her imaging showed markedly dilated lateral ventricles with pneumoventricle, and a widened anterior fontanelle. Her defect was sutured, and she was treated with appropriate antibiotics. She was discharged once infection resolved, but eventually expired at home.

**Discussion:** The second is a 3-year-old male who presented with leaking CSF from a cutaneous rupture at the anterior border of the posterior fontanelle. His head circumference was 92cm, with a 4-centimeter scalp defect exposing the underlying ventricle. The defect was sutured at the emergency room. His family refused further diagnostics and treatment, and opted to bring him home, and was lost to follow-up.

**Results:** Only a few case reports on spontaneous cutaneous rupture are available to guide its treatment, with none coming from the Philippines. Our cases establish the local incidence of this underreported complication and demonstrate that simple suturing and antibiotic treatment do not lead to desirable outcomes.

**Keywords:** Hydrocephalus, Spontaneous Rupture, Philippines, Case Report

## Introduction

Pediatric congenital hydrocephalus is one of the most common reasons for neurosurgical consultation. More than 160,000 new cases of congenital hydrocephalus are diagnosed annually worldwide. Majority are seen in low-to-middle income countries (LMICs)<sup>1</sup>. Patients in the Philippines encounter social and economic challenges to accessing prenatal and neurosurgical care. The COVID-19 pandemic brought delays to the management of severe hydrocephalus infants. A rare complication of this is spontaneous cutaneous rupture leading to cerebrospinal fluid (CSF) leakage. Only a few publications have been written on this phenomenon, mostly coming from LMICs<sup>1, 2, 3, 4, 6, 8</sup>. Moreover, no account of this phenomenon in the Philippines has been published as of this writing.

## Case Presentation

We present two cases of severe hydrocephalus with delayed management, ultimately causing spontaneous rupture and CSF leak. Written consent in using the patient's case were obtained from the parents of the patient.

### Case 1

The first case was a 9-month old female born full term to a 30 year-old mother. This was her second pregnancy, with her first being delivered full-term normally. A prenatal ultrasound was done at the 5th month of pregnancy, which showed normal fetal development. Upon birth at a local hospital, she was noted to have a head circumference of 39cm (>99th percentile) and subsequently admitted at their Neonatal Intensive Care Unit (NICU) for neonatal pneumonia and congenital hydrocephalus. No neurosurgical consultation was done; she was then discharged after two weeks.

In the interim, her head circumference increased, prompting consultation at our institution's online platform. Here she was advised to admission for ventriculoperitoneal shunt insertion. At this time the COVID-19 pandemic emerged, causing further delays.

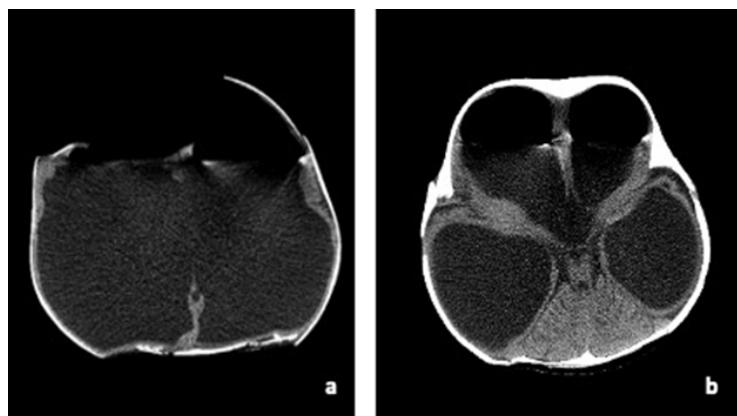
At 9 months, she presented with sudden CSF leakage through a pinpoint cutaneous rupture with associated febrile episodes and was rushed to our emergency room.

On examination, she was awake, with good activity, and briskly isocoric pupils. Her head circumference was 85cm (>99th percentile). The anterior fontanelle was widely open and sunken, and had a pinpoint opening at its right frontal area, with egress of CSF. CSF was taken for analysis through aspiration from this opening which revealed ventriculitis (*Klebsiella pneumoniae*). The opening was dressed using sterile gauze moistened with saline solution, and covered using Tegaderm. This opening progressively increased in size, ultimately reaching 5cm in its widest diameter. The defect was difficult to close without cutting through the thinned-out skin, but was eventually apposed with vertical mattress sutures (Figure 1).



*Fig. 1 Head of patient PP post-suturing showing sunken anterior and posterior fontanelles, with thin skin overlying the everted bony borders. The cranial sutures are also noted unfused further everting the bony fontanelle border.*

Plain cranial CT-scan done showed a widened and depressed anterior fontanelle. Cranial sutures were unfused. The lateral and third ventricles were prominently dilated with pneumoventricle seen in the bilateral frontal horns, with thinning of the frontal, parietal, and occipital cerebral cortices (Figure 2).



*Fig. 2: Select cuts of the plain cranial computed tomography scan done after suturing. Note the thinned out cortex around the dilated lateral ventricles (a), and small fourth ventricle (b).*

Parents were apprised of his poor prognosis. He was treated with appropriate antibiotics for the ventriculitis. Patient was noted to be a poor surgical candidate and was sent home once cleared of the infection, with confirmation through CSF analysis.

On follow-up via teleconsult, patient had expired at home a month after discharge from this institution.

## Case 2

The second case is a 3 year-old male born pre-term to a 29 year-old mother. The patient was the third pregnancy, with the first two being delivered normally at full term. The mother had monthly prenatal check-ups, with intake of folic acid and prenatal vitamins. Prenatal ultrasound was allegedly done at 13 weeks age of gestation, which showed the patient to have congenital hydrocephalus, and cleft lip and palate. No maternal illnesses and radiation exposures were noted.

He was delivered via primary cesarean section at a local hospital. No record of his head circumference at birth was available. They were referred to a neurosurgeon for further management of the hydrocephalus. At three months old, they were offered insertion of a ventriculoperitoneal shunt. However, due to fears of doing brain surgery on their child, the parents refused, and he was discharged a few days after.

In the interim, there was progressive increase in head circumference. At 2 years, his parents eventually consulted at our institution for neurological surgery. However, the COVID-19 pandemic emerged, and his parents opted against admission.

At 3 years, a skin ulceration developed at his occiput, which eventually ruptured, resulting in the sudden egress of CSF, prompting consult at our emergency. No antenatal and postnatal records were available upon presentation to our institution. On examination, he was awake, with decreased activity, pupils isocoric with sunsetting, and a head circumference of 92cm (>99th percentile). Anterior and posterior fontanelles were widely dilated and sunken. A 10 x 4 cm ulceration at the occipital portion of the posterior fontanelle was actively leaking CSF. The cutaneous rupture was sutured, preventing further CSF leakage (Figure 3). After suturing, parents refused any further imaging or ancillaries, and treatment. His parents opted to bring him home, and was lost to follow-up.



*Fig. 3: Thinned out skin overlying the posterior fontanelle with ulceration sutured to further prevent CSF leakage*

## Discussion

The annual incidence of hydrocephalus globally was estimated to reach more than 380,000 cases, and more than 40% of these are congenital in etiology<sup>1</sup>. Most new cases came from the African region, with Southeast Asian and the Latin American regions with second and third most numbers seen—all LMICs. However, majority of published works about the burden of this disease are found in high-income countries, where the burden is lowest<sup>1</sup>.

Delays in the management of severe congenital hydrocephalus cause progressive expansion of the ventricles, thinning of the cerebral and non-fusion of the cranial bones and closure of fontanelles. In its later stages, progression of the ventriculomegaly attenuates the dura and overlying skin, ultimately causing the rare complication of spontaneous cutaneous rupture leading to leakage of CSF<sup>2,3,4</sup>.

Ventricular walls rupture most commonly at the occipital and frontal horns—regions of least resistance due to the relative thinness of the cortices in these lobes<sup>1,3,5</sup>. The sudden relief of high intracranial pressure by this rupture causes depression of the open fontanelles. The non-union of the cranium accompanied by the relief of pressure causes expansion of fontanelle borders. These changes create what other have referred to as an “alien-head deformity”<sup>3,6</sup>. The connection to the external environment predisposes the patient to ventriculitis and sepsis<sup>2,3,6</sup>.

We presented two cases with similar details: both patients were not seen by a neurosurgeon immediately after birth, and eventually consulted during the height of the pandemic. The restrictions brought about by the pandemic for non-urgent and non-emergent cases, such as congenital hydrocephalus, hampered their access to advanced healthcare. Furthermore, resources were channeled towards the COVID response, putting less focus on hydrocephalus. In both cases, the parents sought consultation at the emergency department due to the spontaneous cutaneous rupture and CSF leakage. Finally, both had an “alien-head deformity”, with sinking of the fontanelles and eversion of the cranium at the fontanelles.

Because of the paucity of evidence, no guidelines for the management of patients with cutaneous rupture of severe hydrocephalus have been published and remains a challenge to neurosurgeons who encounter them. Many questions remain unanswered: does any surgery beyond simple suturing offer any benefit, which patients will benefit from surgery, and what form of surgery is appropriate?

Only a few publications have been written on this phenomenon<sup>2,3,4,6,7,8</sup>. Patients ranged from 3 months to as old as 3 years of age. The head circumference was documented in three of these cases, and ranged from 45cm to 73cm.<sup>8</sup> Five proceeded with CSF diversion, with two of these also underwent cranioplasty for correction of the cranial deformity. Of the eight patients in the literature, there were three recorded deaths (one following surgery and two that were conservatively-managed), while no long-term follow-up on the other cases was noted.

Spontaneous cutaneous rupture occurs exclusively in patients with untreated severe hydrocephalus. A recent study compared early CSF diversion against CSF diversion with cranial vault reduction cranioplasty in patients with severe congenital hydrocephalus<sup>9</sup>. They found that combining cranioplasty with CSF shunting in the neonatal period can simplify treatment and result in acceptable cosmesis. However, none of the patients included in this previous study had a spontaneous cutaneous rupture, and the benefits of cranial vault reduction surgery for treating or preventing this complication is unclear.

The patients we presented had head circumferences greater than any other published cases (85cm and 92cm). Both were managed conservatively, primarily eliminating the connection between the internal central nervous system and the external environment by cutaneous closure. Antibiotics were given when possible to treat for ventriculitis. Despite this, the first case presented expired after being discharged from the institution, while the second was lost to follow-up but likely also expired. This demonstrates how simple closure through suturing and antibiotics treatment does not lead to desirable outcomes. In the Philippines, there are undoubtedly many other undocumented cases of severe hydrocephalus with cutaneous rupture that do not reach the attention of neurosurgeons. Future studies can explore the benefit of combining CSF diversion with cranial vault reduction surgery in these unfortunate patients.

## Conclusion

We have presented two cases of severe congenital hydrocephalus with delayed surgical management that resulted in spontaneous cutaneous rupture of the ventricles and CSF leakage. This rare complication increases the risk for CNS infections and mortality. The most appropriate treatment for these patients is not yet clear, but simple suturing and antibiotics alone led to poor outcomes. A combination of CSF diversion and cranial vault reduction surgery may be beneficial, but the most effective intervention is early detection of hydrocephalus and prevention of rupture.

## Conflict of Interest

None declared.

## References

1. Dewan, M., Rattani, A., Rania, Mekary., et al. (2018). Global hydrocephalus epidemiology and incidence: systematic review and meta-analysis. *Journal of Neurosurgery*, 2018; 130(4). Accessed on 29 July 2021. <https://doi.org/10.3171/2017.10.JNS17439>.
2. Katiyar, V., Garg, K., Doddamani, R., Singh, P. K., Singh, M., & Chandra, P. S. (2020). Spontaneous external rupture of hydrocephalus after fontanelle closure: a case report and review of literature. *Child's nervous system : ChNS : official journal of the International Society for Pediatric Neurosurgery*, 2020; 37(6): 2069–2073. Accessed on 9 July 2021. <https://doi.org/10.1007/s00381-020-04915-w>.
3. Tripathi, M., Padwal, A., Shinde Pandit, B., et al. (2014). Unusual complication of neglected hydrocephalus: spontaneous rupture with growing skull fracture. *Pediatric Neurosurgery*, 2014–15;50:281–285. Accessed on 12 January 2022. <https://doi.org/10.1159/000438795>.
4. Ezechukwu, C., Egbonu, I., Chukwuka, J., et al. (2002). Spontaneous rupture of a massive hydrocephalus resulting in head collapse. *Nigerian Journal of Clinical Practice*, 2002; 5(2): 148-149. Accessed on 12 January 2022.
5. Peña, A., Bolton, M., Whitehouse, H., et al. (1999). Effects of brain ventricular shape on periventricular biomechanics: a finite-element analysis. *Neurosurgery*, 1999; 45(1):107-16; discussion 116-8. Accessed on 12 January 2022. doi: 10.1097/00006123-199907000-00026.
6. Gothwal, S., Meena, K., & Meena, K.K. (2021). Spontaneous rupture of a neglected huge hydrocephalus. *Indian Journal of Pediatrics*, 2021; 88: 1145–1146. Accessed on 11 January 2022. <https://doi.org/10.1007/s12098-020-03648-7>.
7. Oyemolade, T., & Balogun, J. (2019). Spontaneous rupture: a rare complication of neglected hydrocephalus. *Child's Nervous System*, 2019; 35:2029–2031. Accessed on 3 February 2022. doi:10.1007/s00381-019-04213-0.
8. Moghtaderi, A., Rahimi-Mvaghar, V., Safdari, M. (2005). Spontaneous brain rupture: a complication of untreated hydrocephalus. *Clinical Neurology and Neurosurgery*, 108: 48-51. Accessed on 3 February 2022.
9. Iyer, R., Carey, C., Rottgers, A., et al. (2018). Early postnatal cranial vault reduction and fixation surgery for severe hydrocephalic macrocephaly. *Journal of Neurosurgery – Pediatrics*, 2018; 21:486-495. Accessed on 9 July 2021. doi: 10.3171/2017.11.PEDS17173.

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