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Pediatric extreme hydrocephalus after shunting: preliminary findings of long-term follow-up

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Abstract

Introduction: *Pediatric extreme hydrocephalus (PEH) is a complex condition with uncertain prognostic outcomes.* **Objectives:** *In this study, we analyze the long-term change in cerebral parenchymal thickness of pediatric patients with extreme hydrocephalus after shunting.* **Methods:** *A retrospective observational study was conducted on patients with PEH treated at the General Hospital of Mexico from 2009 to 2016. Cerebral parenchymal thickness was measured in computed tomography studies before surgical intervention, at 5 months, and 4 years post-surgery. The average change in thickness in millimeters was analyzed, and its normalized value was assessed using the Wilcoxon test and the R^2 of a linear function.* **Results:** *Twelve patients, including 5 females and 7 males, were studied. Nine cases were congenital, while the remaining 3 cases were 6 months, 7, and 10-years-old. At 5.2 months PostOp., patients showed an average increase of 160% in cerebral parenchymal thickness, which increased to 270% at 4.3 years. However, 4 cases (33.3%) had an increase of < 10% from the original thickness. One patient at 4.3 years exhibited a 594% increase compared to the baseline. In the congenital cases, pre-operative thickness had a good correlation (R^2 = 0.70) with post-operative thickness, but pre-operative age of patients did not correlate with post-operative thickness (R^2 = 0.03).* **Conclusions:** *In this study, 3 out of 12 cases of PEH showed a long-term increase of > 200% compared to preoperative thickness. Even in cases of extreme hydrocephalus, significant long-term improvement can occur. Rapid treatment of these cases is crucial to increase the likelihood of improvement.*

Keywords: *Extreme hydrocephalus. Pediatric hydrocephalus. Structural neuroplasticity.*

Introduction

Hydrocephalus is one of the most frequent and relevant neurosurgical disorders in both adult and pediatric patients. In the pediatric population, hydrocephalus ranks as the third leading cause of admission to the Neurosurgery Department at the National Institute of Pediatrics in Mexico[1]. Similarly, at the General Hospital of Mexico, congenital hydrocephalus has been one of the leading causes of birth abnormalities[2]. In most cases of pediatric hydrocephalus, there is a moderate to significant accumulation of cerebrospinal fluid (CSF). However, in certain cases, there is a massive accumulation of CSF[3-6]. The compression exerted by the excessive amount of CSF on the brain parenchyma can severely affect brain development. In fact, a greater amount of CSF is expected to have a more severe impact on neuronal tissue, sometimes leading to the absence of cortical tissue in extensive areas[3,7].

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Our research problem statement is that these cases of extreme hydrocephalus pose a unique and complex challenge, especially concerning the management and progression of these patients. Interestingly, to date, there are no descriptions of the long-term effects of surgical treatment on parenchymal thickness in pediatric patients with extreme hydrocephalus. In this study, we examine the chronic changes in brain thickness in pediatric patients with extreme hydrocephalus after undergoing ventriculoperitoneal shunt placement at the Hospital General de México.

Materials and methods

A retrospective, observational, and longitudinal study was conducted using the records of pediatric patients treated at the Hospital General de México "Dr. Eduardo Liceaga."

Subjects

The study included patients of both sexes, aged between 0 (newborns) and 18 years, diagnosed with extreme hydrocephalus and treated with ventriculoperitoneal shunt placement in the pediatric neurosurgery department of the General Hospital of Mexico. Patients with hydrocephalus secondary to tumor-related causes and those without long-term imaging studies (> 3 months post-surgery) were excluded from the study. The study period ranged from September 1, 2009, to January 30, 2016. The present study protocol was submitted for registration to the Research Committee of our hospital.

Retrospective measurements

In cranial computed tomography studies, the axial section at the approximate level of the foramen of Monro was identified; in this section, the transition from the frontal horn to the atrium of the left lateral ventricle was identified. Then, the thickness of the cerebral parenchyma was measured from the pial surface to the ventricular surface (Fig. 1A: dashed line), and the distance between both inner tables was also measured at the same level (Fig. 1: solid double-head line).

Data analysis

The post-surgery parenchymal thickness in each patient's studies was adjusted as a ratio relative to the diameter between the inner tables (Fig. 1A, blue line) and normalized to the pre-surgery thickness. Values were expressed as a percentage relative to the pre-surgical baseline thickness. These normalized percentage data were grouped into two time points (5.2 months and 4.3 years), corresponding to the average times at which control imaging studies were conducted for operated patients. In cases of congenital hydrocephalus, the normalized parenchymal thickness data at 5.2 months were plotted against the age at treatment. In these same cases, parenchymal thickness in millimeters was also plotted against pre-surgery thickness in millimeters. In both cases, the R^2 value of a linear function was calculated to assess the degree of correlation. Grouped data were expressed as a percentage or as mean ± standard deviation. The Wilcoxon test was used to determine if the average normalized values at 5.2 months and 4.3 years differed. A $p < 0.05$ was considered statistically significant. Data collection and analysis were conducted in Excel program.

Results

The present study included 12 cases, 7 male and 5 female patients. Nine cases had congenital hydrocephalus (average age at diagnosis: 11.0 ± 7.6 days), while the remaining three cases were aged 6.9 months, 7.1 years, and 10.7 years, respectively (Fig. 1). The thickness of the cerebral parenchyma in the motor/perimotor region of the left hemisphere was measured in all cases before surgery and after ventriculoperitoneal shunting, at 5-6 months (average: 5.2) and 4-5 years (average: 4.3) post-surgery. At the time of diagnosis, prior to shunting, the cerebral parenchymal thickness was 18.48 ± 12.66 mm. However, when considering only cases of congenital hydrocephalus, the parenchymal thickness was 14.11 ± 10.45 mm. Given the considerable intrinsic variation found in the cases, data were adjusted relative to the diameter between the inner tables and normalized to the pre-surgical baseline for temporal comparison. In the follow-up studies, the cases showed an increase in normalized cerebral parenchymal thickness of 160 ± 61% and 270 ± 128% at 5.3 months and 4.3 years of follow-up, respectively (Figs. 1 and 2). At 5.2 months, 5 patients (41%) exhibited an increase in cerebral parenchymal thickness of > 200%, while in 4 cases (33.3%), there was no significant improvement (< 10% increase). One patient at 4.3 years showed an increase of 594% compared to the initial pre-surgical thickness (Fig. 2).

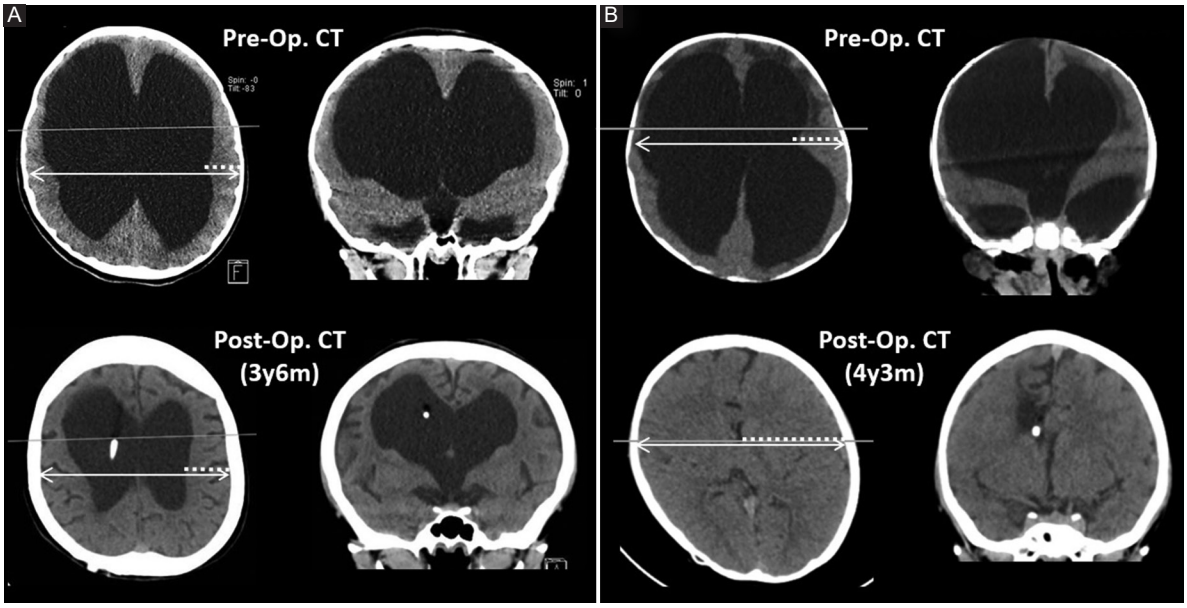

Figure 1. Computed tomography (CT) scans of two cases of extreme hydrocephalus treated with ventriculoperitoneal shunting. **A**: case 2, female, 10 years 9 months of age at the time of diagnosis (upper images) and 3 years 6 months after shunt placement (lower images). **B**: case 4, male, 6 months of age at the time of diagnosis (upper images) and 4 years 3 months after shunting. The horizontal continuous lines represent the level of CT sections; the dashed white lines on the axial images indicate the region where cerebral parenchymal thickness was measured, and the double-head arrow lines show how the intracranial diameter was measured at the same level.

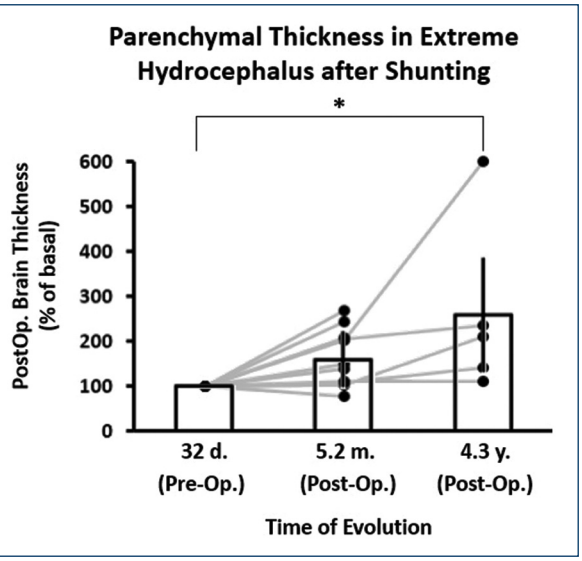

Figure 2. Temporal course graph of cerebral parenchymal thickness of shunted patients. Data are adjusted and normalized to the preoperative thickness (left column) of each case. Central and right columns with error bars represent post-operative mean values ± standard deviation, dots represent individual patients, *p < 0.05. mean value ± standard deviation.

Considerable variability was found in the development of cerebral parenchymal thickness and since 75% (n = 9) of the cases studied were congenital hydrocephalus, we analyzed whether there was a relationship between the time of evolution (neonatal age) and the change in parenchymal thickness at 5 months in these congenital cases. We found that the change in parenchymal thickness did not show a proportional relationship with the age of surgical treatment (Fig. 3A, $R^2 = 0.03$). However, when plotted against the pre-surgical parenchymal thickness, we observed that a greater pre-shunting parenchymal thickness was associated with a greater tendency to have increased cerebral parenchymal thickness at 5 months post-surgery (Fig. 3B, $R^2 = 0.70$).

All cases were successfully managed for hydrocephalus; however, shunt system revision was performed in three cases. Two revisions were due to shunt system dysfunction, and other one was due to CSF leakage into the subgaleal space at the trephination site. Another case experienced CSF infection, requiring externalization of the system, antibiotic treatment, and replacement with a new shunt.

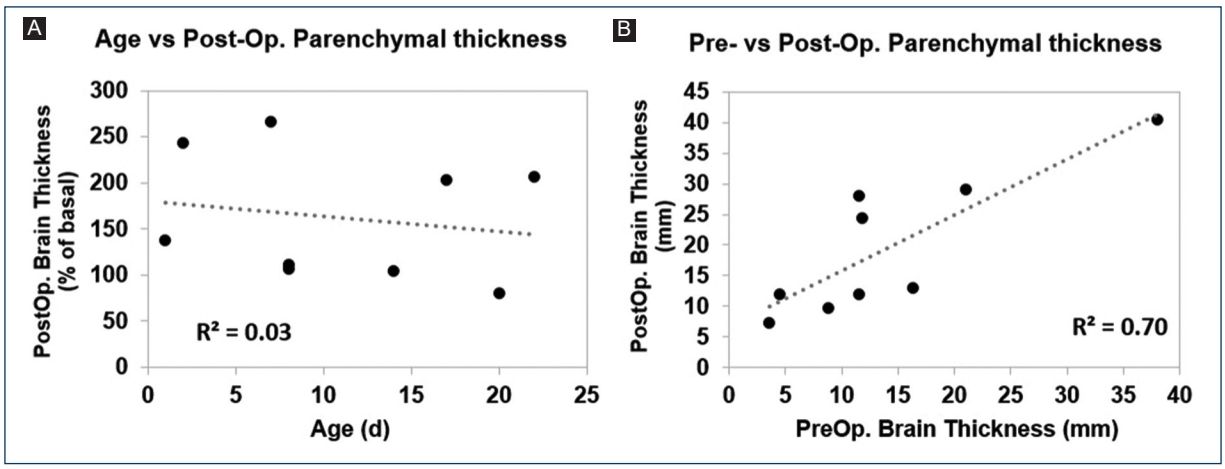

Figure 3. Correlations of postoperative cerebral parenchymal thickness in congenital extreme hydrocephalus. **A**: age vs. normalized parenchymal thickness at 5.2 months post-surgery. **B**: pre-operative parenchymal thickness versus post-operative thickness at 5.2 months. Note the R^2 value of 0.70 in the second graph.

Discussion

In the present study, we observed that even in extreme cases of hydrocephalus with severe compression of neuronal tissue, significant long-term improvement and recovery in cerebral parenchymal thickness can occur.

The first reports of extreme hydrocephalus (also known as maximum or massive hydrocephalus) were made in the 1960s[3,4]. In several cases, patients were not surgically treated as clinical findings suggested a poor prognosis, which was associated with intrauterine onset of hydrocephalus. Thus, cerebral tissue thickness was a decisive factor in determining whether a patient underwent surgical treatment or estimating their prognosis. In contrast, in our institution all the patients were shunted independently of the initial cerebral thickness. It has been shown that cortical mantle thickness did not show a deterministic correlation with the neurological status of patients[3,4]. Currently, it is known that extreme hydrocephalus *per se* is not a contraindication for intervention[5,7,8], but the presence of comorbidities (myelomeningocele and malnutrition) and delayed CSF diversion negatively influence the prognosis. In the present work, there are not patients with comorbidities, except malnutrition, and the delayed CSF shunt was only considered in the congenital cases. Interestingly, the data suggest that age is not directly or intrinsically important (Fig. 3A), but rather depends on the severity of hydrocephalus and the degree of parenchymal compression (Fig. 3B). An important consideration for diagnosis of these cases is whether there is extreme hydrocephalus or anencephaly. Sutton et al. (1980) observed that the latter cases show minimal occipital parenchyma and absence of EEG activity, which is useful for the diagnostics[7]. Although we did not conduct EEG studies on our patients, in hindsight, we observed that all treated patients were cases of PEH, as they all showed an increase in cerebral parenchyma. All cases in this report were treated with ventriculoperitoneal shunting; however, there are more than one alternative surgical treatments that can be effective for certain cases. Shitsama et al. (2014) reported stabilization of macrocephaly in 40% of cases of extreme hydrocephalus or anencephaly treated with endoscopic choroidal plexus coagulation[5].

Interestingly, the recovery in cerebral parenchymal thickness even in cases of massive hydrocephalus demonstrates the capacity for recovery and plasticity of the nervous system in certain cases. While neuron density and number are important for proper neurodevelopment, cases with massive CSF accumulation have been observed in which the brain can generate almost normal phenotype and behavior[9,10]. The cases reported serve as a reminder of the difficulty in managing these patients. Moreover, despite the challenges and possible complications of surgical treatment, like those observed in the patients in the present study, it is central to comprehensively treat these patients with a multidisciplinary management.

Finally, it is important to mention the limitations of this study: (a) it is an observational and retrospective study, (b) due to the nature and frequency of the condition, few cases were studied, and (c) in the 4-year follow-up,

some cases were lost because follow-up was not continued at our hospital. Given the above, it is necessary to have new data and case series in the future, preferably in controlled prospective studies.

Conclusions

In the present study, 25% of the patients with extreme hydrocephalus showed long-term recovery (> 200%) in cerebral parenchymal thickness. Timely diagnosis and immediate treatment are crucial in these cases.

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Conflicts of interest

The authors declare that they have no conflicts of interest.

Ethical disclosures

Protection of human and animal subjects. The authors declare that no experiments were performed on humans or animals for this study.

Confidentiality of data. The authors declare that they have followed the protocols of their work center on the publication of patient data.

Right to privacy and informed consent. The authors had to obtain the approval of the Ethics Committee for the analysis and publication of routinely obtained clinical data. The informed consent of the patients was not required because this was a retrospective observational study.

Use of artificial intelligence for generating text. The authors declare that they have not used any type of generative artificial intelligence for the writing of this manuscript, nor for the creation of images, graphics, tables, or their corresponding captions.

References

1. Marhx-Bracho A. Hidrocefalia. In: Matilde RG, Rueda-Franco F, editors. Manual de Neurología y Neurocirugía Pediátricas. 1st ed. Mexico City: Alfil; 2016. p. 24.
2. Ortiz-Almeralla MR, Flores-Fragoso GF, Cardiel-Marmolejo LE, Luna-Rojas CL. Frecuencia de malformaciones congenitas en el área de neonatologia del Hospital general de México. Rev Mex Pediatr. 2003;70:128-31.
3. Yashon D, Jane JA, Sugar O. The course of severe untreated infantile hydrocephalus. Prognostic significance of the cerebral mantle. J Neurosurg. 1965;23:509-16.
4. Lorber J. The results of early treatment of extreme hydrocephalus. Dev Med Child Neurol. 1968;Suppl 16:21-9.
5. Shitsama S, Wittayanakorn N, Okechi H, Albright AL. Choroid plexus coagulation in infants with extreme hydrocephalus or hydranencephaly: clinical article. J Neurosurg Pediatr. 2014;14:55-7.
6. Di Rocco C, Iannelli A. Poor outcome of bilateral congenital choroid plexus papillomas with extreme hydrocephalus. Eur Neurol. 1997;37:33-7.
7. Sutton LN, Bruce DA, Shut L. Hydranencephaly versus maximal hydrocephalus: an important clinical distinction. Neurosurgery. 1980;6:35-8.
8. Ray C, Mobley J, Thompson M, Nagy L. Hydranencephaly: considering prolonged survival and treatment by endoscopic choroid plexus coagulation. Turk Neurosurg. 2015;25:788-92.
9. Alders GL, Minuzzi L, Sarin S, Frey BN, Hall GB, Samaan Z. Volumetric MRI analysis of a case of severe ventriculomegaly. Front Hum Neurosci. 2018;12:495.
10. Ferris CF, Cai X, Qiao J, Switzer B, Baun J, Morrison T, et al. Life without a brain: neuroradiological and behavioral evidence of neuroplasticity necessary to sustain brain function in the face of severe hydrocephalus. Sci Rep. 2019;9:16479.