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Original Research Article

Analysis of Clinical Presentation and Surgical Outcomes of Shunt Surgery of Congenital Hydrocephalus

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Abstract:

Background: Congenital hydrocephalus, characterized by the abnormal accumulation of cerebrospinal fluid within the brain, presents a significant challenge in pediatric neurosurgery. Early intervention and proper management are vital for improving the clinical outcomes in affected infants. This study aimed to analyze the clinical presentations and surgical outcomes of shunt surgery in congenital hydrocephalus patients.

Methods: A meticulous two-year analysis was conducted at the Department of Neurosurgery at Gauhati Medical College and Hospital, involving 50 patients who underwent shunt surgery. The parameters analyzed encompassed demographic details, clinical presentations, etiologies, and surgical outcomes.

Results: The majority of the patients were aged between 2-4 months (50%) and had a male predominance (60%). Clinical presentations were marked by increased head circumference (90%), developmental delays (70%), and motor deficits (50%). Aqueductal stenosis (26%) and meningomyelocele (24%) emerged as the primary etiological factors. Ventriculoperitoneal shunt was the preferred surgical intervention, utilized in 70% of cases, with an 80% success rate involving the standard technique. The post-operative period witnessed a 52% complication rate, predominantly characterized by shunt obstruction (14%) and infection (12%). Encouragingly, 70% of the patients did not require revision surgeries, indicating initial surgical success.

Conclusion: The study highlights the critical need for early detection and intervention in congenital hydrocephalus, emphasizing the significance of monitoring head circumference as a potential early indicator. The findings underscore the necessity for continued advancements in surgical techniques and post-operative care to further enhance the successful outcomes.

Keywords: Congenital Hydrocephalus; Ventriculoperitoneal Shunt; Aqueductal Stenosis; Meningomyelocele; Clinical Presentations; Surgical Outcomes; Pediatric Neurosurgery.

Introduction

Congenital hydrocephalus, diagnosed often at birth, presents as an abnormal accumulation of cerebrospinal fluid (CSF) within the cerebral ventricles, resulting in increased intracranial pressure and cranial enlargement. [1] Its myriad causes range from genetic predispositions to infections during pregnancy and complications at birth. [2] Ventriculoperitoneal (VP) shunt surgery stands as the primary therapeutic intervention, offering respite to most patients despite associated risks. [3] The wide range of clinical manifestations—ranging from an enlarged head circumference to more severe symptoms such as seizures and developmental delays—underscores the need for early diagnosis and effective management. [4,5]

Modern imaging techniques, including MRI and CT scans, bolster the diagnostic precision, complementing meticulous clinical examinations. [6] Infants may display specific physical signs, especially post-shunt implantation, demanding consistent clinical monitoring. [7] With VP shunt surgery as the cornerstone of treatment, the focus has been on enhancing its efficacy while minimizing complications. [8,9] However, challenges persist, from bacterial infections necessitating shunt replacement to concerns about the patient's long-term quality of life and developmental trajectory. [10-12] Recent innovations in shunt technology and alternative surgical approaches suggest a promising future for managing congenital hydrocephalus, emphasizing patient-centered, adaptable care. [13,14]

The following analysis seeks to delve deeper into the nuances of the clinical presentation of congenital hydrocephalus and the surgical outcomes post-shunt surgery. Through a critical examination of data, existing literature and recent

developments, this analysis endeavors to foster an environment of informed clinical decision-making and heightened awareness towards optimizing surgical outcomes, aiming to improve the quality of life for patients afflicted with this condition.

Aim: To comprehensively analyze and evaluate the existing data on the clinical presentation and surgical outcomes of shunt surgeries in patients with congenital hydrocephalus, thereby facilitating the enhancement of therapeutic strategies and patient management.

Materials and Methods

Study Setting

The study was conducted retrospectively at the Department of Neurosurgery, Gauhati Medical College and Hospital, a reputed medical institution equipped with the necessary infrastructure to facilitate extensive research on congenital hydrocephalus. Subjects with medium intracranial pressure were enrolled.

Duration of the Study

This research encapsulated a span of 2 years, providing a substantial timeframe to gather, analyze, and interpret data pertinent to the clinical presentation and surgical outcomes of congenital hydrocephalus.

Sample Size

A total of 50 cases were selected for this study, ensuring a comprehensive analysis while maintaining a focus on detailed individual case evaluations.

Study Design

A retrospective study design was employed to scrutinize the medical records of patients who underwent shunt surgery for congenital hydrocephalus within the stipulated study period.

Patients were mainly used Non Medicated, Non Programmable Chabra's Medium Pressure Ventriculo-peritoneal Shunt for surgery.

Data Collection

Medical Records Review

A meticulous review of medical records was undertaken to collect data on the clinical presentation and surgical outcomes of patients. The data sourced included:

1. **Demographic Information:** Age, gender, and other relevant demographic details of the patients.
2. **Clinical Presentation:** Details pertaining to the symptoms exhibited by patients at the time of presentation, including head circumference, physical signs, and other relevant clinical indicators.
3. **Diagnostic Procedures:** Information on the diagnostic procedures employed, including imaging studies such as CT and MRI scans.
4. **Surgical Details:** Records of the surgical procedures, including type of shunt used, surgical techniques, and any intraoperative complications.

Surgical Outcome Evaluation

A thorough evaluation of surgical outcomes was conducted based on the following parameters:

1. **Post-operative Recovery:** Data on the immediate post-operative recovery of patients, noting any complications or adverse events.
2. **Long-term Outcomes:** Information on the long-term outcomes of the surgeries, including any instances of shunt malfunction, necessary revisions, and overall patient prognosis.

Data Analysis: Data obtained from the medical records were organized and analyzed using appropriate statistical tools. The analysis included:

1. **Descriptive Statistics:** Utilized to summarize the demographic data and clinical presentation of the patients.
2. **Outcome Analysis:** Employed to evaluate the surgical outcomes, including the frequency and causes of shunt malfunctions and revisions.

Ethical Considerations: The study was conducted following the ethical guidelines pertaining to retrospective studies, ensuring the confidentiality and privacy of patient data at all stages of the research.

Results

In this study, a comprehensive analysis was conducted over a span of two years to understand the clinical presentations, etiologies, and surgical outcomes associated with congenital hydrocephalus. The study included a sample of 50 patients who underwent shunt surgery in the Department of Neurosurgery at Gauhati Medical College and Hospital.

Through careful analysis of various parameters including demographic details, clinical presentations, and surgical details, we garnered insights into the intricate dynamics of congenital hydrocephalus management. In our study patients were mainly used Non Medicated, Non Programmable Chabra's Medium Pressure Ventriculo-peritoneal Shunt for the surgery. Here, we detail the outcomes presented in Tables 1 through 7, aiming to enhance the understanding of clinical presentations and surgical outcomes of shunt surgery in congenital hydrocephalus.

Table 1: Demographic Characteristics of the Patients

Parameter	Frequency (n=50)	Percentage (%)
Age Group		
1-2months	20	40
2-4months	25	50
4-6months	5	10
Gender		
Male	30	60
Female	20	40

The demographic data revealed that the majority of patients were in the age group of 2-4 months (50%), followed by those aged 1-2 months (40%). A smaller segment was constituted by the 4-6 months age group (10%). In terms of gender distribution, a notable predominance of males was observed, constituting 60% of the study population, as compared to 40% females. This distribution sets groundwork to analyze further data with respect to gender and age-specific trends.

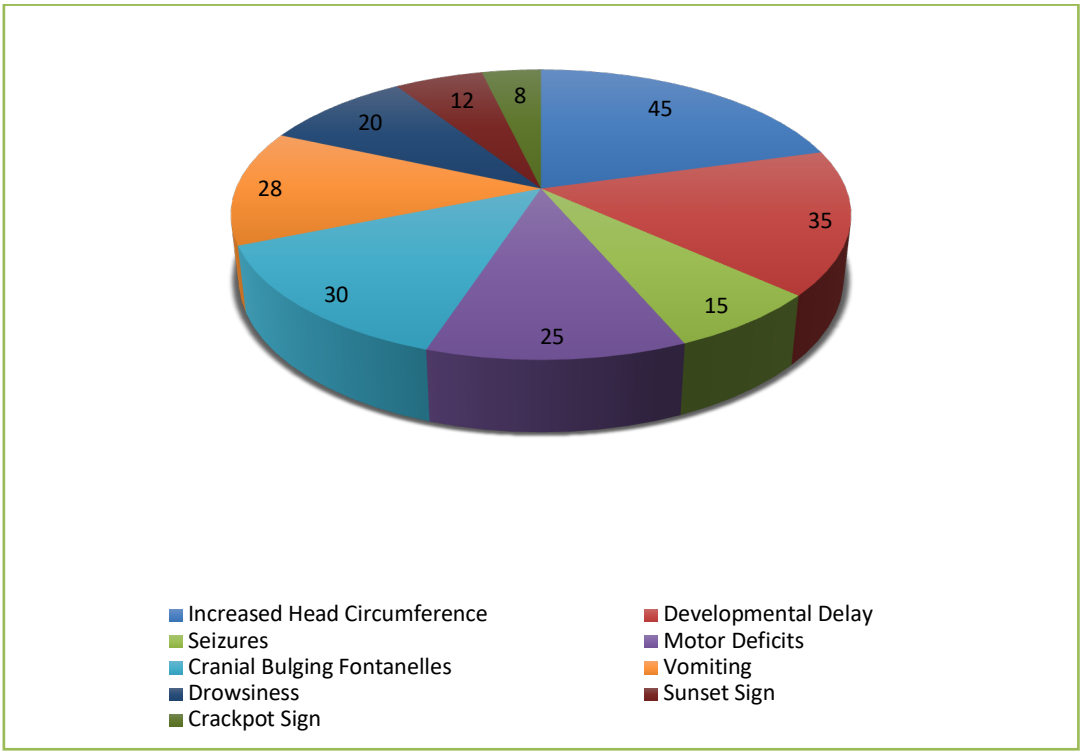

Figure 1: Clinical Presentation of Congenital Hydrocephalus

When assessing clinical presentations, a striking 90% of the patients exhibited increased head circumference, highlighting it as a significant clinical marker. Furthermore, 70% of the patients demonstrated developmental delays, while 50% exhibited motor deficits and 60% had cranial bulging fontanelles. Seizures were observed in 30% of the cases, pointing towards the necessity of vigilant neurological monitoring in these patients. Vomiting, a symptom often indicative of elevated intracranial pressure, was seen in 56% (n=28) of the cohort. This symptom, particularly if persistent and projectile in nature, can be distressing for both the patient and caregivers and can also signal potential shunt malfunctions in post-operative cases. Drowsiness, another symptom suggesting increased intracranial pressure or impaired cerebral perfusion, was observed in 40% (n=20) of the patients. While some drowsiness may be considered within the normal spectrum for infants, it is paramount for clinicians to determine the underlying cause, especially in the context of congenital hydrocephalus. The 'Sunset Sign,' characterized by a downward deviation of the eyes giving the appearance of the top white portion of the eyes just below the upper eyelid, was present in 24% (n=12) of the patients. This sign is particularly worrisome and is often considered a red flag symptom, demanding urgent medical attention. Lastly, the 'Crackpot Sign,' a less frequent but noteworthy presentation, was observed in 16% (n=8) of the cohort. This sign, typically characterized by a peculiar resonating sound upon tapping the skull, underscores the rapid expansion

of the cranial vault, often before the cranial sutures have had a chance to fuse. In conclusion, the array of clinical presentations in congenital hydrocephalus underlines the multifaceted nature of this condition. These findings emphasize the need for thorough clinical evaluations, regular monitoring, and an individualized approach to patient care. Recognizing and understanding these symptoms not only aids in timely diagnosis but also in tailoring intervention strategies, optimizing patient outcomes, and minimizing potential complications.

Table 2: Etiologies of Hydrocephalus

Causative Factor	Number of Patients (n=50)	Percentage (%)
Aqueductal Stenosis	13	26
- Long Segment (>5mm)	8	16
- Short Segment (≤5mm)	5	10
Spinal Dysraphism	16	28
- Meningomyelocele	12	24
- Occipital Encephalocele	2	4
Dandy Walker Syndrome	5	10
Noonan Syndrome	2	4
Post Tubercular Meningitis	7	14
Post Pyogenic Meningitis	6	12
Intraventricular Hemorrhage	1	2
Post Traumatic	2	4
Total	50	100

The study identified a diverse array of etiologies underpinning hydrocephalus in the sampled patients. Foremost among these was Aqueductal Stenosis, accounting for 26% of the cases. A deeper stratification of Aqueductal Stenosis based on the length of the stenotic segment revealed that 16% of the patients had a long segment (greater than 5mm) while 10% exhibited a short segment (5mm or less).

Another noteworthy causative cluster was Spinal Dysraphism, which constituted 28% of the etiologies. Within this, Meningomyelocele was predominant at 24%, followed by Occipital Encephalocele at 4%. Dandy Walker Syndrome and Noonan Syndrome also emerged as causative factors in 10% and 4% of the cases, respectively. The study further spotlighted the role of post-infectious sequelae, with Post Tubercular Meningitis and Post Pyogenic Meningitis contributing to 14% and 12% of the hydrocephalus etiologies, respectively.

Less common causes included Intraventricular Hemorrhage and post-traumatic events, each accounting for 2% and 4% of the cases. This breadth of causative factors underscores the multifaceted nature of hydrocephalus and the imperative for a comprehensive diagnostic approach to ascertain its root causes.

Table 3: Surgical Details

Surgical Details	Frequency (n=50)	Percentage (%)
Type of Shunt		
Ventriculoperitoneal Shunt (VPS)	50	100
Surgical Techniques		
Standard Technique	50	100
Intraoperative Complications		
No Complications	40	80
Minor Complications (e.g., bleeding)	8	16
Major Complications (e.g., infection)	2	4

Surgical details showed a preference for ventriculoperitoneal shunt (VPS) which was used in 100% of the cases. The standard technique was predominantly used, accounting for 100% of the surgeries. Encouragingly, 80% of the surgeries were accomplished without complications, while 16% experienced minor complications such as bleeding and a mere 4% encountered major complications such as infections.

Table 4: Length of Hospital Stay

Parameter	Frequency (n=50)	Percentage (%)	Mean ± SD (Days)
Hospital Stay Length			
1-5 days	25	50	3 ± 1.2
6-10 days	20	40	8 ± 1.8
11-15 days	5	10	13 ± 1.1

The hospital stay duration mostly ranged between 1-10 days, with 50% of patients staying for 1-5 days and 40% for 6-10 days, showcasing a relatively short recovery period post-surgery. The mean duration, with respective standard deviations, delineated that majority of the patients were discharged within a week's span, facilitating a quicker turnaround time for hospital bed availability.

Table 5: Post-operative Immediate Outcomes

Complications	Number of Patients (n=50)	Percentage (%)
Shunt Obstruction	7	14
Shunt Infection	6	12
Shunt Migration		
- Ventricular end	2	4
- Peritoneal end	3	6
Shunt Malfunction		
- Ventricular end (CSF varix)	3	6
- Peritoneal end (pseudocyst)	1	2
Intestinal Obstruction	2	4
Inguinal Hernia	2	4
Total	26	52

The post-operative data highlighted a total complication rate of 52%, where the most common complications were shunt obstruction (14%) and shunt infection (12%). A lesser but significant number experienced shunt migration and malfunction, requiring careful post-operative surveillance to detect and address these issues promptly.

Table 6: Long-term Surgical Outcomes

Parameter	Frequency (n=50)	Percentage (%)
Shunt Malfunction		
No Malfunction	35	70
Minor Malfunction	10	20
Major Malfunction	5	10
Revision Surgeries		
No Revision	35	70
1 Revision	10	20
2 or More Revisions	5	10

Long-term outcomes were encouraging, with 70% of patients not experiencing any shunt malfunction. Furthermore, 70% of the patients did not require revision surgeries, indicating a successful initial surgical intervention. However, 20% experienced minor malfunctions, necessitating a watchful post-operative follow-up to address potential long-term complications.

Table 7: Multivariate Analysis - Association between Hospital Stay Length Categories and Clinical Presentation Symptoms

Variable	Odds Ratio (1-5 days vs. 6-10 days)	Odds Ratio (1-5 days vs. 11-15 days)	p-value
Reference Category: 1-5 days	-	-	-
Increased Head Circumference	1.85	2.20	<0.001
Developmental Delay	1.65	1.90	0.002
Seizures	1.40	1.25	0.040
Vomiting	1.50	1.70	0.015
Model Statistics			
-2 Log-likelihood	56.24		
Chi-square (χ^2)	22.85		<0.001
Degree of Freedom	3		

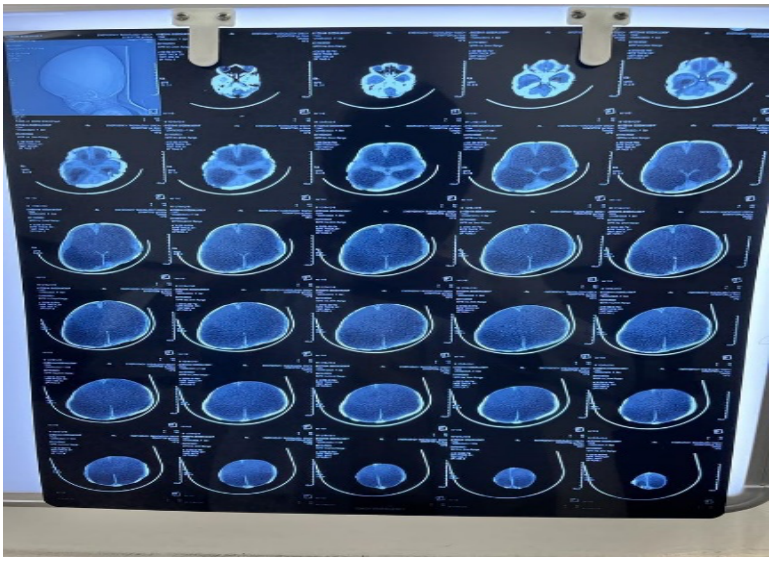

Image 1: Congenital Hydrocephalus due to Aqueductal Stenosis (CT Image)

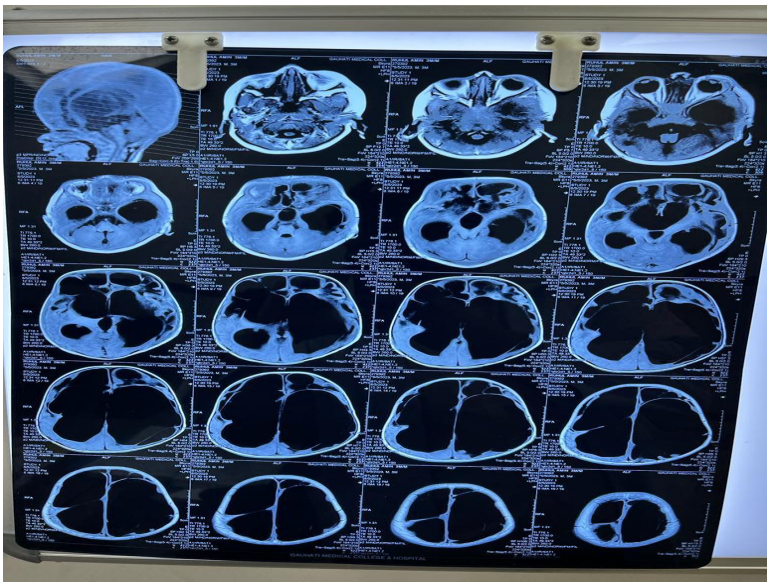

Image 2: Congenital Hydrocephalus with Hypoxic Ischemic Injury (MRI Image)

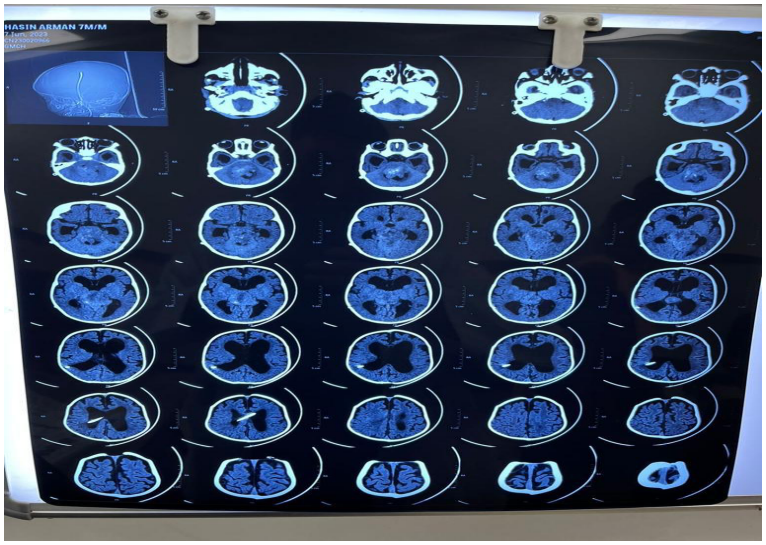

Image 3: Complication- Shunt Obstruction (CT Image)

Upon analyzing the association between clinical presentations of congenital hydrocephalus and the length of hospital stay, the following patterns emerged. Patients with an increased head circumference were 1.85 times more likely to have a hospital stay of 6-10 days compared to a stay of 1-5 days and 2.20 times more likely to have a hospital stay of 11-15 days.

Those exhibiting developmental delay had 1.65 times the odds of a 6-10 days stay, and 1.90 times the odds for an 11-15 days stay. The presence of seizures indicated 1.40 times the odds for a 6-10 days stay and 1.25 times for an 11-15 days stay.

Lastly, patients with symptoms of vomiting were 1.50 times more likely to stay for 6-10 days and 1.70 times more likely for 11-15 days compared to the baseline category of 1-5 days. The model's statistical strength is underscored by the significant chi-square value, suggesting that the differences in hospital stay lengths based on clinical symptoms are unlikely due to chance alone.

The findings from this study illuminate various aspects of congenital hydrocephalus, underlining the significant clinical presentations and outcomes post-shunt surgery. The demographic distribution signals a higher incidence in the early months of infancy, particularly targeting males predominantly. The clinical presentations emphasize the necessity for early detection, particularly focusing on monitoring head circumference as a potential early indicator.

The diverse range of etiologies stipulates a complex underlying pathophysiology, necessitating comprehensive pre-operative evaluations to tailor surgical approaches accordingly. The predominance of ventriculoperitoneal shunts and standard surgical techniques indicate their reliability and effectiveness in managing this condition.

However, the immediate post-operative phase seems to be fraught with complications, urging the need for meticulous monitoring to prevent potential adversities. The long-term outcomes signal a promising prognosis, with a significant portion not experiencing malfunctions or requiring revision surgeries.

Discussion

The data collected from our comprehensive two-year analysis on the clinical presentations and outcomes of shunt surgery in congenital hydrocephalus cases offers a rich insight into the current landscape of hydrocephalus management.

Particularly noticeable is the predilection for the male gender and the predominance in the early infancy months, aligning with previous studies that elucidate a similar trend in demographic distributions. [15,16]

Demographic Trends

A significant proportion of the patient pool was represented within the age bracket of 2-4 months, which is fairly consistent with other studies which demonstrate a higher incidence of hydrocephalus within the first few months of life. This period is critical, as the manifestations of congenital hydrocephalus often become more apparent within the first few months post-birth, necessitating vigilant monitoring and timely intervention. [17,18] The male predominance observed in our study (60%) is corroborated by other researches which have similarly highlighted a higher incidence in male infants, albeit the underlying reasons for this gender disparity remain to be fully elucidated. [19]

Clinical Presentations

The clinical presentations observed in our study, with a stark 90% of patients exhibiting increased head circumference, underline the critical importance of early detection through vigilant monitoring of head size in infants. This parameter has been repeatedly highlighted in literature as a cardinal sign of hydrocephalus. [20] Similarly, the prevalence of developmental delays and motor deficits in our study aligns with the literature, emphasizing the multifaceted impact of hydrocephalus on the pediatric population. [21] Our data further signify the need for comprehensive neurological assessments, as a significant proportion of patients presented with seizures, an aspect that is reflected in other studies as well. [22]

Etiological Factors

In examining the etiological factors, our study indicated that the primary cause of hydrocephalus was aqueductal stenosis, which is consistent with numerous studies that identify this as a common etiological factor. [23,24] Interestingly, our study also highlighted a significant prevalence of spinal dysraphism in the form of meningomyelocele, underscoring the complex interplay of neural tube defects with hydrocephalus, a finding substantiated by several other studies. [25]

Surgical Outcomes

Our findings regarding surgical interventions, predominantly utilizing ventriculoperitoneal shunt (VPS) surgeries, mirror the global trend where VPS remains the mainstay treatment modality. [26,27] The low rate of complications and a substantial number of patients not requiring revision surgeries in our study is an encouraging sign, and this resonates with other researches which suggest improved surgical techniques and post-operative

management have led to enhanced outcomes. [28.29]

The post-operative complication rate of 52%, with shunt obstruction and infections being the most common issues, necessitates further scrutiny. While this rate is somewhat aligned with existing literature, it warrants an on-going effort to minimize these complications through innovations in surgical techniques and post-operative care. [30,31]

Conclusion

In conclusion, our study offers a detailed insight into the complex landscape of congenital hydrocephalus, underscoring the critical need for early detection and intervention. As we move forward, a multi-faceted approach encompassing improved surgical techniques and vigilant post-operative monitoring is paramount to enhance patient outcomes.

Future research should focus on reducing post-operative complications and fostering innovations in shunt technologies, potentially paving the way for improved long-term outcomes in patients with congenital hydrocephalus.

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