Original Article

Study of Neurodevelopmental Outcome in Patients with Non-tumoral Hydrocephalus with Shunt Surgery Done in Infancy

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INTRODUCTION

Hydrocephalus (HC) is a common central nervous system disorder in childhood. It represents a diverse group of conditions that result from impaired circulation and absorption of cerebrospinal fluid (CSF) or, in rare circumstances, from increased production of CSF by choroid plexus papilloma.

It causes excess of CSF accumulation within the ventricular system and cisterns of the brain, leading to dilatation of the cerebral ventricular system, increased intracranial pressure (ICP) with corresponding compression effects on the brain parenchyma and associated structures such as cranial nerves. It may lead to blindness due to atrophy of the optic nerve, memory impairment, deterioration of intellectual capacity, and difficulties in movement such as gait disturbances.¹

Background: Hydrocephalus (HC) is a common neurological disorder presenting in infancy, with a myriad of etiologies requiring early neurosurgical intervention.

Objective: To study neurodevelopmental outcome in patients with HC with shunt surgery done in infancy.

Materials and Methods: This was an observational retrospective cohort study of 50 pediatric patients (2 years to 16 years of age). These patients were diagnosed with HC and were operated on with ventriculo-peritoneal shunt (VP shunt) insertion in infancy (did not include patients with brain tumors) and then later following in the neurology outpatient department (OPD). Clinical records and neurodevelopmental assessment (intelligence quotient [IQ]/development quotient [DQ] and vision and hearing assessment) were reviewed.

Results: Only 50% of the patients with congenital HC were diagnosed at birth, which included patients who had been diagnosed antenatally and they had lesser complications and better intellectual outcome (P = 0.12), compared with those who presented later with HC. Patient-related factors such as etiology of HC, antenatal diagnosis, and requirement of shunt revisions had poor correlation with neurodevelopmental outcome. Patients with late postoperative complications had significantly poor neurodevelopmental outcome (P ≤ 0.001). Patients with post-meningitis HC required a significantly higher number of shunt revisions than patients with other causes (P = 0.04).

Conclusion: Better neurodevelopmental outcome depends on early diagnosis and early referral for the management than the cause of HC. Regular head circumference monitoring is the most feasible and sensitive screening tool for early pickup. Larger studies are needed for accurate prognostication.

Keywords: Infantile VP shunt outcome, neurodevelopmental outcome of pediatric hydrocephalus, pediatric hydrocephalus outcome

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Diagnosis has been made easier with various neuroimaging techniques such as magnetic resonance imaging (MRI), which adds to the already existing transfontanellar ultrasound (TFU), and the computerized tomography (CT) scan.\[^{2}\]

HC can be classified according to (1) timing of presentation (congenital, acquired), (2) pathogenesis (communicative or obstructive), and (3) etiology (aqueductal stenosis, post-meningitic etc.). Clinical features and modality of treatments may vary according to the type of HC.\[^{3,4}\]

Pediatric HC is a surgical disease. VP shunt is the gold standard of treatment. Endoscopic third ventriculostomy is rapidly gaining preference as an alternative. Medium-pressure and low-pressure shunts are chosen based on intraoperative manometry and anterior fontanelle at the time of surgery. With present-day standard of care, most patients with HC will survive but if left untreated, case fatality is high.\[^{5,6}\] The sequelae among long-term survivors are frequent and often severe.\[^{7}\] Because of the multiplicity of causes of HC, associated diseases, complications of treatment, and the inherent complexity of the patient population, reliable data on outcomes are difficult to obtain. The outcome of patients with HC has been the subject of many studies, but most of the studies have limitations that are due to either narrow focus or conflicting results.\[^{8,9}\] Despite its importance, it has not been sufficiently studied in developing countries such as India.

In our study, we have reviewed and analyzed the past records of patients with non-tumoral HC who were presented, diagnosed, and shunted in infancy. Neurodevelopmental assessment, including assessment of vision and hearing, was performed. An attempt was made to determine the role of each factor with the ultimate neurodevelopmental outcome of the patient in a systematic way to draw useful conclusions in the management of HC in early life.

**Materials and Methods**

This was an observational retrospective cohort study that was conducted in children in the age group of 2 years to 16 years. Ethics committee approval was taken before commencement of the study. Patients who were operated for non-tumoral HC in infancy and were followed up in the pediatric neurology OPD were enrolled in the study if they fulfilled the study criteria (informed written consent taken appropriately). The study period was two years (from June 2016 to July 2018). Privacy and anonymity of patients were maintained throughout the study.

All past records were reviewed for clinical details. Records of head circumference, surgical details and their complications, revision of shunt, laboratory, and imaging details were noted. Their detailed neurodevelopmental assessment was done by using the modified Denver development scale test (DDST) for an assessment of DQ for less than six years and the Wechsler Intelligence Scale for Children (WISC) for an assessment of IQ for the 6- to 16-year age group. Ophthalnic assessment (by an age appropriate test) and auditory evaluation (by brainstem evoked response audiometry/audiometry) were also done in each patient. All the patient-related factors were correlated with their impact on neurodevelopmental outcome.

**Results**

**Epidemiology and clinical presentation**

In our study, 50 children were enrolled: 60% \((n = 30)\) of them were between two and five years of age, and 72% were males; 66% \((n = 33)\) cases were congenital ones, and the rest were acquired HC; and 9 \((27\%)\) of 33 patients with congenital HC were antenatally diagnosed. HC due to aqueductal stenosis was the most common etiology \((30\%, n = 15)\) followed by meningitis \((24\%, n = 12)\) and meningomyelocele/arachnoid cyst \((12\%, n = 6)\) each. Most were an obstructive variety of HC \((88\%, n = 44)\). From one month to six months, the most common feature was the age of presentation \((36\%); the increasing head size was the most common presenting feature \((88\%); and this was followed by signs of raised intracranial pressure such as vomiting \((30\%); seizures \((20\%); and irritability \((10\%)\) [Table 1].

**Surgical results**

Overall, 14% \((n = 7)\) patients initially required a reservoir to drain CSF temporarily before shunt; 58% \((n = 29)\) patients did not require any shunt revision; 22% \((n = 11)\) patients required it once; and 20% \((n = 10)\) patients required more than one shunt revision. Shunt block \((52\%, n = 11)\) was the most common indication of revision followed by shunt discontinuation \((38\%, n = 8)\) followed by shunt infection \((9.5\%, n = 2)\). Overall, 50% of patients who required shunt revision were cases of post-meningitis (including tuberculosis) HC; 10% \((n = 5)\) of patients had postoperative hemiparesis and another 10% \((n = 5)\) had shunt-related infection postoperatively; and 80% of patients were free from early postoperative complications. Epilepsy \((36\%, n = 18)\) was the most common late complication followed by intellectual disability, intestinal obstruction, and over-drainage of ventricles.
Neurodevelopmental outcome

Overall, 21 out of 50 (42%) patients showed normal development with no disability (ID/DQ >85%). On retrospective analysis, it was found that all of them were detected at birth, had fewer surgical complications and revisions (mean revisions of 0.38 compared with overall average of 0.72, \( P = 0.176 \)), and had no tone and gait abnormalities. Overall, 36% (\( n = 18 \)) of patients had mild to moderate intellectual disability, and 22% (\( n = 11 \)) had severe to profound intellectual disability. Overall, 80% of patients had normal vision, 12% (\( n = 6 \)) had suboptimal vision, and 8% (\( n = 4 \)) had no vision (all these cases had aqueductal stenosis); 90% (\( n = 45 \)) of patients had normal hearing, 8% (\( n = 4 \)) had mild to moderate hearing loss (all post-aqueductal stenosis), and 1 patient had profound hearing loss (post-meningitis). Overall, 42% (\( n = 21 \)) of the total patients were nonambulatory, which included all patients with meningo(myelo)cele [\( n = 6 \)], 50% [\( n = 3 \)] patients with arachnoid cyst, 47% [\( n = 7 \)] patients with post-meningitis HC, and 33% [\( n = 5 \)] patients with HC due to aqueductal stenosis; 20% (\( n = 10 \)) patients had an unsteady gait.

DISCUSSION

The management of pediatric HC is both medical and surgical. Medical management includes management of acute symptoms of increased ICP. Surgical management remains the mainstay of treatment. With an improvement in standards of care, most patients with HC will survive; however, death from HC still exists, and the sequelae among long-term survivors are frequent and often severe. Impaired mobility (e.g., cerebral palsy), impaired cognition (intellectual disability, behavior problems), sensory deficits (vision, hearing), and epilepsy are the most important specific concerns in the early life of these patients. Timely decision making and appropriate intervention leads to favorable outcomes. Further prognosis depends on multiple factors.

Because of the multiplicity of causes of HC, complications of treatment, and the inherent complexity of the patient population, reliable data on outcomes are difficult to obtain. In previous studies, multiple combinations of factors have been studied but the data are not enough to determine the outcome at the time of initial diagnosis. There are hardly any studies from the Indian literature and so conclusive guidelines are lacking.

In our study, congenital HC (66%) was found to be more prevalent than acquired HC. Overall, aqueductal stenosis (30%) was the most common cause of HC followed by post-meningitis (24%) and then HC due to spinal dysraphism (12%) was the third most common in the study. Other causes were arachnoid cyst, Dandy

| Sl. no. | Variable | Groups            | \( N \) | Mean ± SD | \( P \) value |
|--------|----------|-------------------|--------|-----------|--------------|
| 1      | Type of HC | Congenital        | 33     | 73 ± 34   | 0.25         |
|        |          | Acquired          | 17     | 63 ± 30   |              |
| 2      | Underlying cause of HC | 9 causes | 50 | 78 ± 36 | 0.36 |
| 3      | Antenatal diagnosis | Yes | 9 | 78 ± 36 | 0.36 |
|        |          | No                | 41     | 68 ± 32   |              |
| 4      | Presentation of congenital HC | At birth | 13 | 85 ± 31 | 0.12 |
|        |          | Later             | 20     | 66 ± 35   |              |
| 5      | Duration of hospital stay during first shunt | <30 days | 41 | 74 ± 32 | 0.03 |
|        |          | >30 days          | 9      | 50 ± 30   |              |
| 6      | Side of shunt | Right-sided shunt | 43 | 69 ± 32 | 0.46 |
|        |          | Left-sided shunt  | 7      | 77 ± 38   |              |
| 7      | Type of shunt | Medium-pressure shunt | 42 | 71 ± 32 | 0.54 |
|        |          | Low-pressure shunt | 8      | 63 ± 37   |              |
| 8      | No. of revisions required | 0 | 29 | 71 | 0.75 |
|        |          | 1                 | 11     | 72        |              |
|        |          | 2                 | 8      | 61        |              |
|        |          | ≥3                | 2      | 63.5      |              |
| 9      | Presence of late postoperative complications | Present | 24 | 55 ± 31 | 0.00078 |
|        |          | Absent            | 26     | 84 ± 28   |              |
| 10     | Birth weight | SGA | 20 | 58 ± 36 | 0.04 |
|        |          | AGA               | 30     | 77 ± 28   |              |
| 11     | Association between type of HC and revision requirement | Post-meningitic HC | 12 | 1.33 ± 1.6 | 0.04 |
|        |          | Other causes of HC | 38     | 0.52 ± 0.8 |              |
Walker malformation, Arnold Chiary malformation, and craniostenosis in decreasing frequency. HC secondary to post-intraventricular hemorrhage was the least common cause of HC requiring a shunt in infancy. All these trends are consistent with previous studies such as those by Mouafo Tambo et al.,[10] in which they found aqueductal stenosis in 31.43% cases.

The mean DQ/IQ of patients with congenital variety was found to be 73.0 ± 68.5 more than those with acquired HC variety (63.7 ± 59.0), which was possibly due to early detection and intervention as well as the association with lesser comorbidities; however, difference was statistically insignificant (P = 0.25), likely due to the small sample size in our study. In his Indian series, Upadhyaya did not find any correlation between the nature of HC and age at operation with intellectual outcome, which is consistent with our study.[11] The subtypes of HC such as Dandy Walker malformation (100 ± 0) and meningoencephalocele (82.5 ± 23.9) were found to have a better mean DQ/IQ compared with other subtypes, but these differences were found to be statistically insignificant (P = 0.75) due to their small sample size. This reveals that outcome depends on cause but more on management.

Only 27% of proven congenital HC was diagnosed antenatally on routine antenatal sonography and referred, which is suggestive of the poor sensitivity and subjective variation of the antenatal screening program. Patients with congenital HC that presented at birth had a comparatively uneventful neonatal course compared with those who presented later, as they had neonatal complications such as sepsis, respiratory distress etc. and a few of them (25%) had a prolonged NICU stay of more than 6 weeks. They also had less immediate/late postsurgical complications than patients with congenital HC who presented later on. The average DQ/IQ of those with congenital HC presented at birth was found to be higher (83.15 ± 62.1) than that of those who presented later (66.4 ± 70.68), although this difference was not very statistically significant (P = 0.12). It emphasizes the importance of antenatal screening for better outcomes.

Apart from those antenatally diagnosed, 18% of patients with congenital HC were diagnosed at birth and 15% by the first month of life. However, 40% of patients were diagnosed later between 2 and 11 months of life. This indicates that congenital HC may exhibit symptoms/signs at a later age or may be missed in the early part of life, which can be picked up on regular head circumference monitoring.

Associated factors such as preterm delivery and low birth weight added to the comorbidities of patients requiring prolonged intensive care, further worsening the neurodevelopmental outcome evidently and emphasizing the importance of good general neonatal care for better outcomes. Those who are SGA had better DQ/IQ (58 ± 36, n = 20) compared with those who are AGA (77 ± 28, n = 30), which was statistically significant. Persson et al.[12] concluded that being born very preterm and with HC that is already overt at birth involves the highest risk of a poor outcome.

In the study, it was found that the lesser the gap between parents/primary physician noticing the HC and referral to the tertiary center, the better was the prognosis. Although earlier referral does not mean immediate surgical intervention, it may lead to proper follow-up and timely intervention. Thus, it is evidenced here that cases of HC should be referred to appropriate centers immediately after detection. However, the decision of surgery may differ based on the neurosurgeon’s discretion. Correlation between the duration of HC (duration between the age of onset and the age of shunt surgery) and final outcome (IQ/DQ) was found to be weak, with a Pearson correlation coefficient value (R value) = 0.031 and P value 0.83.

In the study, 88% of patients presented with increasing head circumference (including all 100% of congenital HC and 70% of acquired HC). Most of them (57%) were asymptomatic but the remaining showed symptoms of mechanical compression of brain parenchyma such as vomiting/posturing/alterned sensorium (congenital > acquired) or features of sepsis such as fever/seizure (acquired > congenital). These findings again give utmost importance to regular head circumference monitoring for early diagnosis and, thus, improving outcome.

A reservoir was placed in 14% of cases before the shunt. Shunt side (right or left side) and type of shunt (medium- or low-pressure shunt) did not have any implication on final neurodevelopmental outcome.

Only 42% of patients required a revision of shunt, 22% required it once, 16% required two revisions, and 8% required it ≥3 times. Shunt functioning rate was 90% at one year, 82% at two years, and 74% at 5 years.

Shunt block was the most common cause of revision (52.3%) followed by discontinuation (breakage). Displacement and shunt infection were the least common cause of revision. Patients with post-meningitis HC required maximum revisions (mean = 1.33 ± 1.67) compared with other causes all together (mean = 0.52 ± 0.82), which was found to be statistically significant (P = 0.04, U = 305.50). Antenatal diagnosis, type and side of shunt, congenital/acquired type of disease, and
timing of diagnosis do not seem to have any statistically significant effect on the average number of revisions.

Overall, 18% of patients experienced immediate complications such as hemiparesis (10%), shunt-related infections (6%), and pseudocyst formation at the abdominal end (2%). Those patients with immediate complications also had late complications.

Overall, 48% of patients had late sequelae, with epilepsy being the most common (36%), intellectual deficit (8%) and hemiparesis (6%) commonly at the opposite side of the shunt. Over-drainage of ventricles and intestinal obstruction due to adhesions were very uncommon complications. All those patients with late complications had statistically significant lower DQ/IQ compared with patients without complications.

The overall complication rates were found to be low (48%) in our study group compared with previous studies.

The overall average duration of stay was 14 days in total (pre- and postsurgery). Those with hospital stay <30 days were found to have higher DQ/IQ of 74.12 ± 63.76 than those with hospital stay >30 days (50.44 ± 63.08), which was statistically significant ($P = 0.03$).

In the study, 40% patients had DQ/IQ ≥90%. All patients with high DQ/IQ were found to be detected at birth. They also had a low frequency of late complications and lesser number of revisions (0.38 compared with overall average of 0.72) ($P = 0.176$). No significant association was found between DQ/IQ with gender, type of HC, antenatal diagnosis, side of shunt, and number of revisions.

Overall, 80% of patients had normal vision, 12% of patients had poor vision; and 8% were totally blind; the majority of these cases of abnormal vision had aqueductal stenosis (72%) and arachnoid cyst (18%). Similarly, 90% had normal hearing, 8% had moderate to severe hearing loss, and 2% had profound hearing loss. Among patients with hearing loss, 40% were post-meningitis sequelae and the remaining cases were due to aqueductal stenosis and arachnoid cyst. All these patients had a statistically significant low DQ/IQ, indicating that severe CNS injury due to disease and deficiency of sensory input were causes of poor intellectual outcome.

Tone/power/reflexes were very well correlated with gait outcomes and DQ/IQ. This shows that HC has a negative effect on both motor functions and cognitive abilities in a similar proportion based on the severity of disease.

With the present-day standard of care, baseline imaging is done before shunting, for ascertaining the cause, better positioning of the shunt, prognostication of complications, and outcome.

It is challenging to measure and prognosticate the outcome of all types of pediatric HC, as it requires a larger sample size and long-term follow-up, which are difficult to obtain. Studies at a larger scale are needed.

**Conclusions**

Pediatric HC is a common CNS disorder in developing countries, and its neurodevelopmental outcome is multifactorial. The majority of congenital HC are missed by antenatal scans. Half of the congenital HC presented at birth and they had lesser complications and better intellectual outcome. Regular checkup and keeping track of head circumference can be rewarding in early diagnoses of HC. Early detection and referral to a higher center is of utmost importance in reducing the perioperative complications and thus improving the outcome. Aqueductal stenosis is the most common cause of congenital HC, and it has poor influence on visual outcome. Post-meningitis HC is the most common cause of acquired HC and it has a higher number of revisions and poor influence on hearing outcome compared with the other subtypes. Better perinatal care reduces the comorbidities, and better perioperative care improves outcome. Larger sample size and long-term follow-up are needed for precise prognostication and for further improvement of pediatric HC care.

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**Ethical approval**

The study was conducted after getting ethical approval from the Ethics Committee, Seth GSMC and KEM Hospital, Parel, Mumbai 12.

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**Conflicts of interest**

There are no conflicts of interest.

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