Quality of Life in Individuals Surgically Treated for Congenital Hydrocephalus During Infancy: A Single-Institution Experience

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BACKGROUND: **Congenital hydrocephalus (CH) is a frequently encountered birth anomaly that can hinder long-term neurologic maturity and social well-being of affected children. This study was undertaken to assess quality of life (QOL) 10–15 years after surgical treatment for primary CH during infancy at a tertiary care hospital in a developing country.**

METHODS: **This retrospective cohort study included individuals who presented to Aga Khan University Hospital, Karachi, Pakistan, between 1995 and 2005 at <1 year old and underwent surgery for primary CH. The Hydrocephalus Outcome Questionnaire was used to assess outcomes with respect to QOL.**

RESULTS: **Of 118 patients, 90 patients participated in the study. Mean age at first admission was 6.2 months. Mean length of follow-up was 5.4 years. Of these, 28 patients had died after surgery. Shunt infection ($P = 0.012$) and delayed milestones ($P = 0.003$) were found to be statistically significant factors affecting mortality in the patients who died. The mean overall health score was 0.67 ± 0.30. Age <6 months at the time of first surgery was a poor predictor of overall health on the Hydrocephalus Outcome Questionnaire ($P = 0.039$).**

CONCLUSIONS: **In our analysis, we assessed the QOL associated with CH. We hope that these results will provide insight for future prospective work with the ultimate goal of improving long-term QOL in children with CH.**

Key words
- Congenital
- Hydrocephalus
- Hydrocephalus Outcome Questionnaire (HOQ)
- Quality of life (QOL)

Abbreviations and Acronyms
CH: Congenital hydrocephalus
HOQ: Hydrocephalus Outcome Questionnaire
QOL: Quality of life

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Supplementary digital content available online.

Citation: World Neurosurg. (2017) 101:247-253.
http://dx.doi.org/10.1016/j.wneu.2017.01.107

Journal homepage: www.WORLDNEUROSURGERY.org

Available online: www.sciencedirect.com

INTRODUCTION

Congenital hydrocephalus (CH) is a common birth disorder.[1] Increased intracranial pressure in affected children makes them prone to future seizure disorders, cognitive decline, and mental retardation.[2,3] A previous study reported that approximately one third of patients with CH had seizure disorders, and approximately 40% had an IQ of <70.[2,3] Lindquist et al.[4] reported considerable difficulty in cognition even in children with hydrocephalus who had a normal or a near-normal IQ.

Quality of life (QOL) reflects a subjective assessment of emotional, cognitive, and physical health and social well-being in individuals.[5] The perceived impact of a health-impairing condition on QOL is different depending on whether the disease is congenital, acquired early in life, or develops at a later stage.[4] Kulkarni et al.,[6] using a self-designed questionnaire, the Hydrocephalus Outcome Questionnaire (HOQ), reported that parents perceived frequent seizures and shunt-related complications as some of the predictors of poor QOL in children with CH. Gupta et al.[7] studied patients >20 years old who had had early-onset hydrocephalus and reported depression in more than two thirds of them.

Pakistan is a developing country with an estimated population of 191 million, a per capita gross domestic product of approximately 6% of the world's average, and an average literacy rate of approximately 55%. CH is the most common neurologic congenital defect in Pakistan, and most children with CH are treated surgically.[8,9] However, CH involves long-term follow-up and is associated with numerous possible complications. Rashid et al.[10] reported a poor surgical outcome in children treated for pediatric hydrocephalus in Pakistan. Considering the prevalent poverty as well as the lack of well-developed social rehabilitation programs in Pakistan, evaluation of QOL in patients treated for CH requires the identification, survey, and analysis of such patients in our health care system.

The purpose of this study is to examine the management and prognosis of patients treated for CH in a resource-limited setting such as ours. Up to this time, there have been no data available from Pakistan to investigate and address QOL in patients who survive after being treated for CH during their infancy. We believe this study will help highlight the social burden of this disease and increase its awareness at a national level, initiating analysis on a larger scale and ultimately leading to the establishment of more rehabilitation centers in Pakistan. Surgical intervention and long-term follow-up to prevent complications are not enough for the proper care of these patients. Accepting that premise, this study assessed the QOL of patients 10—15 years after they were treated surgically for primary CH during their infancy at a tertiary care hospital in a developing country.

MATERIALS AND METHODS

Approval from the ethics board was obtained before this retrospective cohort study was initiated. All patients who had been treated surgically for primary CH during the period 1995—2005 at Aga Khan University Hospital, Karachi, Pakistan, were included in the study. Of 118 patients identified, 90 fulfilled the inclusion criteria and were selected for this study. There were 28 patients excluded because of either incomplete or missing records ($n = 8$), being operated on in another hospital ($n = 11$), or not giving consent to participate in the study ($n = 9$). Data were retrieved through patient files using International Classification of Diseases, 9th Revision, code 742.3. Patients who had hydrocephalus secondary to meningitis, tumor, or subarachnoid/intraventricular hemorrhage were excluded. The HOQ, a standardized 51-item questionnaire employing the domains of physical, social-emotional, and cognitive health to measure QOL, was used with permission from Kulkarni,[11] who developed and tested this questionnaire.

DATA COLLECTION

We used Kulkarni's HOQ questionnaire, which has been tested for reliability and construct validity and has demonstrated excellent test-retest reliability (0.93, 95% confidence interval, 0.88—0.96), interrater reliability (0.88, 95% confidence interval, 0.79—0.93), and internal consistency (Cronbach's α 0.94).[6] The response of each item is on a 5-point scale, given a score from 0 to 4. For all questions, the following values are entered manually for the various response options:

Not at all true: 0

A little true: 1

Somewhat true: 2

Quite a bit true: 3

Very true: 4

The scores of the items are summed and then divided by the highest possible summed score to provide a final score on a 0.0 to 1.0 metric.

$$\begin{aligned}
\text{Physical Health score} &= \left(\sum[\text{scores of items } 1-15]\right) \div 60\\
\text{Social} - \text{Emotional Health score} &= \left(\sum[\text{scores of items } 16-39]\right) \div 96\\
\text{Cognitive Health score} &= \left(\sum[\text{scores of items } 40-51]\right) \div 48\\
\text{Overall Health score} &= \left(\sum[\text{scores of items } 1-51]\right) \div 204
\end{aligned}$$

Scores of 0.00 ("worse health status") and 1.00 ("better health status") were used for HOQ score interpretation.

Information regarding patient demographics, clinical presentation, and complications of surgery was collected through review of medical records using another self-designed structured questionnaire. Complications were noted for the entire follow-up period. The HOQ was mostly completed by parents of patients and by patients themselves where applicable, with the assistance of a single dedicated neurosurgery resident with at least 2 years of experience. This was done to minimize bias and interobserver variability. Where required, phone calls were made to respondents for the completion of data. Almost all patients were followed through clinics and medical records. However, phone calls were made for the completion of data in some cases because of long distance constraints (e.g., a different city of residence). The data were collected after obtaining consent from the respondent, ensuring protection of the patient's identity and confidentiality during all stages.

Statistical Analysis

Data were analyzed using IBM SPSS Statistics for Windows, Version 21.0 (IBM Corp., Armonk, New York, USA). Continuous data were presented as means with SDs. Categorical data were presented as frequencies and proportions. Nonparametric tests including independent sample Mann-Whitney U test and Kruskal-Wallis test were used to compare continuous data. Several independent variables were tested for possible association with QOL, using the HOQ overall health score as the dependent variable. P value of <0.05 was taken as significant. Univariate analysis was performed on all independent factors hypothesized to indirectly affect QOL parameters. Multivariate analysis using regression analysis of variance was performed on variables that showed statistically significant results on univariate analysis.

RESULTS

Of 90 patients who had undergone the insertion of a ventriculoperitoneal shunt, 40 (44.4%) were male, and 50 (55.6%) were female. Mean age of patients at first admission was 6.2 months; 70 (77%) patients were operated on before 6 months of age. The mean length of hospital stay was 21 days. The mean follow-up time after the surgery was 5.4 years. Complications developed in 56 (62.2%) patients who underwent surgery, whereas 34 (37.8%) patients did not experience any complications. The