

Hydrocephalus: Epidemiology, Etiology & Clinical Outcomes

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ABSTRACT

Background: Hydrocephalus is a condition wherein excess of cerebrospinal fluid (CSF) accumulates within the ventricular system and cisterns of the brain. The aim of this study to evaluate the epidemiology, etiology and clinical outcomes of hydrocephalous patients in paediatrics in PBM hospital Bikaner.

Material & Methods: This is a prospective observational hospital based study conducted in paediatrics and neurosurgery department of PBM hospital Bikaner. All children diagnosed as having hydrocephalus, aged from birth up to 16 years. Clinical examinations were also done.

Results: The present study observed that out 100 cases of hydrocephalus 52 cases had acquired hydrocephalus & 48 cases had congenital. Out of 100 cases, 33% cases present with TBM, 20% had Aq. stenosis, 16% cases had NTD, 8% had pyogenic meningitis. So the most common cause is TBM and children below 2 yrs of age were commonly affected. Males were more commonly affected than females & 30% of cases had hydrocephalous without macrocephaly. Out of 100 cases, 72% were discharge after treatment, 4% were lost to follow up and 24% cases expired and 57% cases had epilepsy. Ophthalmological outcome shows that out of 40 children 30% children had refractive error & out of 100 cases 20% optic

atrophy, 18% had squint, 11% had nystagmus and out of 52 children 58% children shows impaired speech.

Conclusion: Hydrocephalus cases are associated with high mortality and morbidities like epilepsy, ophthalmological and speech problems. Early detection and treatment of hydrocephalus plays crucial role to prevent its complications and mortality.

Key Words: Hydrocephalus, Tuberculosis Meningitis, CSF.

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INTRODUCTION

Hydrocephalus is a condition wherein excess of cerebrospinal fluid (CSF) accumulates within the ventricular system and cisterns of the brain leading to increased intracranial pressure (ICP) and related consequences. This can apparently result from various causes that can affect a fetus, infant, child or adult (Rekate). Summarily, it can be described as an imbalance between production and absorption of CSF.¹ Over production of CSF can also be a cause of hydrocephalus due to choroid plexus tumors, but this is rare (tumors) in clinical practice.

Recently, the role of CNS microcirculation in the absorption of CSF has contributed to the understanding of the pathogenesis of hydrocephalus. As these mechanisms are not still clear, we are forced to follow our understanding and classification based on traditional concepts of CSF circulation.²

The incidence of congenital hydrocephalus is about 0.2–0.5/1000 live births. A higher incidence has been reported in elderly primiparous mothers. It can be associated with a variety of physiological and pathological conditions.^{3,4} Whatever be the etiology, hydrocephalus is further divided into congenital and acquired forms. The etiology of congenital hydrocephalus remains obscure. An inheritable form of aqueductal stenosis has been described in males (X-linked hydrocephalus). The other mechanism of hydrocephalus is over production of CSF, seen in choroid plexus papillomas.⁵

Rekate has classified hydrocephalus based on the CSF flow obstruction. Impaired absorption is another mechanism wherein venous sinus occlusions, vein of Galen malformations and developmental anomalies like craniostenosis with malformation of

the skull base can lead to formation of hydrocephalus. Diseases of the arachnoidal villi can also result in hydrocephalus due to impaired absorption.⁶

Hydrocephalus in patients with TBM could be either communicating or obstructive type, the former being more common.⁷ In both instances, the main cause is the inflammatory exudate occupying the subarachnoid spaces or the ventricular pathways. In the earlier stages of the disease, the thick gelatinous exudates block the subarachnoid spaces in the base of the brain (notably the interpeduncular and ambient cisterns) leading to communicating hydrocephalus.⁸ The exudates lead to a dense scarring of the subarachnoid spaces in the later stages of the disease leading to communicating type of hydrocephalus.

A communicating hydrocephalus can also result from the exudates blocking the arachnoid granulations which prevent the absorption of cerebrospinal fluid (CSF). The inflammation of the choroid plexus and ependyma leads to an overproduction of CSF in the acute phase of the illness, this contributes to the hydrocephalus and raised intracranial pressure.

The obstructive type of hydrocephalus develops when the fourth ventricular outlets are blocked by the exudates or leptomeningeal scar tissue or when there is obstruction of the aqueduct either due to a strangulation of the brain stem by exudates or by a subependymal tuberculoma. Communicating type of hydrocephalus is more common in TBM than the obstructive type. Schoeman et al⁷ found that the hydrocephalus was of the communicating type in 82% of their patients with TBM.

The aim of this study was to evaluate the epidemiology, etiology and clinical outcomes of hydrocephalous patients in paediatrics & neurosurgery department in PBM hospital Bikaner.

MATERIALS & METHODS

This was a prospective observational hospital based study conducted in paediatrics and neurosurgery department of PBM hospital Bikaner.

All children diagnosed as having hydrocephalus, aged from birth up to 16 years. The study was approved by the Institutional Ethical Clearance (IEC) committee, SPMC, Bikaner.

Inclusion Criteria

All admitted children diagnosed as hydrocephalus aged ≤16 years of age.

Exclusion Criteria

Children whose parents denied for consent.

Methods

History of the disease, including developmental history, family history, and duration of the illness. The records of MRI, CT, ultrasound and CSF examination were seen in patients. Clinical examinations was done.

RESULTS

The present study observed that out of 100 cases of hydrocephalus 52 cases had acquired hydrocephalus & 48 cases had congenital (table 1). Out of 100 cases, 33% cases present with TBM, 20% had Aq. stenosis, 16% cases had NTD, 8% had pyogenic meningitis. So the most common cause is TBM (table 2) and children below 2 yrs of age were commonly affected (table 3). Among 100 children, 61% were males and 39% were females. So male: female ratio = 1.5:1. Males were more commonly affected than females (table 4).

In this study showed that 68% cases were weight for age less than -2SD for age & 60 % cases were height less than -2SD for age (table 5) and 70% cases were head circumference more than 2SD for age. 30%of cases have hydrocephalous without macrocephaly (table 6).

Out of 100 cases, 72% were discharge after treatment, 4% were lost to follow up and 24% cases expired (table 7) and 57% cases had epilepsy (table 8). Ophthalmological outcome shows that out of 40 children 30% children had refractive error. Out of 100 cases 20% had optic atrophy, 18% had squint, 11% had nystagmus (table 9) and 58% children shows impaired speech (table 10).

Table 1: Distribution of cases according to pattern of Hydrocephalus

Pattern of hydrocephalus	Number of cases (N=100)
Congenital	48(48%)
Acquired	52(52%)
Total	100(100%)

Table 2: Distributions of cases according to Etiology of Hydrocephalus

Etiology of Hydrocephalus	Number of cases (N=100)
Tubercular meningitis [TBM]	33
Aqueduct stenosis	20
Neural tube defects [NTD]	16
Pyogenic meningitis	8
Arnold chiari malformation	6
Post HIE	5
Tumoral	4
Left cerebellar arachnoid cyst	2
Intracranial haemorrhage [ICH]	2
Dandy walker malformation	2
Mucopolysaccharidosis	1
Craniosynostosis	1
Total	100(100%)

Table 3: Distribution of cases of Hydrocephalus according to Age

Age group	Number of cases (N=100)
<1 month	28(28%)
1month-2 years	41(41%)
2-16 years	31(31%)
Total	100(100%)

Table 4: Distribution of Cases of Hydrocephalus according to Sex

Sex	Number of cases (N=100)
Male	61(61%)
Female	39(39%)
Total	100(100%)

Table 5: Distribution of cases of Hydrocephalus according to Weight & Height for age

Characteristics →	Weight	Height
Age ↓		
<2 SD	68%	60%
>2 SD	32%	40%
Total	100%	100%

Table 6: Distribution of cases of Hydrocephalus according to Head circumference for age

Characteristics →	Head Circumferences
Age ↓	
<2 SD	30%
>2 SD	70%
Total	100%

Table 7: Distribution of cases of Hydrocephalus according to outcome

Outcome	Number of cases (N=100)
Discharged	72(72%)
Lost to follow up	4(4%)
Expired	24(24%)
Total	100(100%)

Table 8: Distribution of cases of Hydrocephalus according to Epilepsy

Outcome	No. of cases
Epilepsy present	57
Epilepsy absent	43
Total	100

Table 9: Distribution of cases of Hydrocephalus according to Ophthalmological Outcome

Ophthalmological Outcome	Number of cases
Squint	18(100)
Nystagmus	11(100)
Refractive error	12(40)*
Optic atrophy	20(100)

*Refractive error could not be assessed in all children because some of them were newborn, comatose, expired.

Table 10: Distribution of cases of Hydrocephalus according to Speech development

Speech	Number of cases (N=52*)
Impaired	30(58%)
Normal	22(42%)

*Out of 100 patients 52 could be assessed for speech and rest of them could not be assessed .

DISCUSSION

The children in this study were of different ages, majority (69%) of them were less than two year and this goes with the findings of a previous study by Aziz et al⁹ and Reem et al.¹⁰ This can be explained by the fact that congenital hydrocephalus and infantile hydrocephalus are usually diagnosed early because of their obvious clinical pattern of presentations i.e. (increase head size, sun setting appearance, dilated scalp veins and bulging fontanelles). Males (61%) outnumbered females i.e. there is slight male predominance and this correlates with the ratio in previous study done in Sudan by Adam et al¹¹, Reem et al¹⁰ and Alebous et al (2012).¹² Although generally it is known that in hydrocephalus there is no sex predominance but there are some cases of aqueduct stenosis that is inherited as sex linked recessive trait and parents have gender bias, so they seek early medical advice for male child.

Concerning the disease characteristics: hydrocephalus in this study was mainly Acquired (52%), out of them (78.9%) were associated with meningitis in which patients with TBM were 63.4%. our results correlate with study of Aziz⁹ in which the acquired cases of hydrocephalus were (76.4%) and the congenital hydrocephalus (23.6%). This was attributed to the epidemic of meningitis in 1993 in Sudan. The majority of children in the study (60%) had stunted growth, with their height less than -2SD for age and 68% of them had weight less than -2SD for age. This could be attributed to the main manifestations of hydrocephalus especially those having increased intra cranial pressure which lead to vomiting and refusal of feed. These results correlates with the previous study by Reem et al.¹⁰

In our study; 70% of children had their HC above 2SD and remaining 30% had their HC less than 2SD, which implies 30% of cases have hydrocephalus without macrocephaly which correlates with previous study done by Reem et al.¹⁰

Hydrocephalus has very poor outcome, the treatment of hydrocephalus requires long-term care and lifelong follow-up. Out of 100 children, 24 children expired, 4 lost to follow up and 72 were discharged. So there is high mortality associated with Hydrocephalus. There is high mortality in Acquired cases (31%) as compared to congenital cases (17%). Children with hydrocephalus often have a convulsive disorder. Out of 100 children 57 had epilepsy. These results correlate with Persson et al.¹³ Epilepsy is more common in acquired type (73%) compared to congenital type (40%) with OR 4.14 and significant P value (0.0007). Hydrocephalus may cause multiple ophthalmological and visual disorders, visual function deficits were identified in majority of children investigated in this study. Out of 100 children 18 had squint, 11 had nystagmus, 20 had optic atrophy. 12 out of 40 child had refractive error. Ophthalmological outcomes are worst in acquired cases compared to congenital. Out of 100 children, 68% shows ophthalmological abnormality. These results are correlates with Persson et al.¹⁴

CONCLUSION

Our study concluded in majority of patient's hydrocephalus is acquired, mostly associated with tubercular meningitis. Hydrocephalus cases are associated with high mortality and morbidities like epilepsy, ophthalmological and speech problems. Early detection and treatment of hydrocephalus plays crucial role to prevent its complications and mortality.

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