Addis Ababa University, College of Health Sciences

School of Medicine, Department of surgery

Neurosurgery Unit

Addis Ababa, Ethiopia

Treatment Outcome of Hydrocephalus among patients with Dandy Walker Malformation managed with ETV at Zewditu Memorial Hospital, from January 2021- May 2023.

NOVEMBER, 2023

ADDIS ABABA, ETHIOPIA

Treatment Outcome of Hydrocephalus among patients with Dandy Walker Malformation managed with ETV at Zewditu Memorial Hospital, from January 2021- May 2023 Addis Ababa, Ethiopia.

Principal Investigator: Dr. Bereket Hailu, MD, Neurosurgical Resident, Neurosurgical Unit, Department of Surgery, Addis Ababa University.

Advisors: - Dr. Kibruyisfaw Zewdie, MD., Assistant Professor of Neurosurgery (Neurosurgery Unit, Department of surgery, AAU, Black Lion Specialized Hospital Addis Ababa, Ethiopia)

- Dr. Yemisirach Bizuneh, MD., Assistant Professor of Neurosurgery (Neurosurgery Unit, Department of surgery, AAU, Black Lion Specialized Hospital Addis Ababa, Ethiopia)

A Research Paper Submitted to Black Lion Specialized Hospital, School of Medicine, Department of Neurosurgery for the partial fulfillment of graduate course in neurosurgery specialty.

Addis Ababa, Ethiopia

November 2023

ACKNOLEGMENT

I would like to extend my heartfelt gratitude to my advisors Dr. Kibruyisfaw Z. and Dr.Yemisirach B. for their continuing support and guidance to develop and conduct this research project, I would also like to extend my appreciation to everyone for their generous support.

TABLE OF CONTENTS

Content Page

ACKNOLEGMENT 3

Lists of Tables 6

List of Figures 7

Acronym and abbreviations 8

Abstract 8

1. Introduction 10

1.1 Background 10

1.2 Statement of the Problem 11

1.3 Justification 11

2. Literature Review 13

3. Objectives 15

3.1 General Objectives 15

3.2 Specific Objectives 15

4. Methods and materials 16

4.1 Study Area 16

4.2 Study period 16

4.3 Study design 16

4.4 Source and study Population 16

4.4.1 Source Population: 16

4.4.2 Study Population: 16

4.5. Eligibility criteria 16

4.5.1 Inclusion criteria 16

4.5.2 Exclusion Criteria 17

4.6 Sample Size 17

4.7Operational Definition 17

4.9 Study Variable 18

4.10 Data Collection tool procedure 18

4.11 Data Quality Assurance 18
4.12 Data Processing and analysis 18
4.13 Ethical Consideration 19
4.14 Dissemination of results 19
5. Results 19
Demographic characteristics 19
Clinical presentation 20
Neuro-imaging Evaluation 21
Complications 21
Treatment Outcome 21
Figure 2 . Kaplan-Meier Survival Plot of Patients with infection vs. patients with no infection. 24
6. Discussion 26
7. Strength and Limitations 28
7.1 Strength 28
7.2 Limitation 28
8. Conclusion and Recommendation 29
9. Reference 30
Annex 31
Questionnaire Tool 31
APPROVAL SHEET 35

Lists of Tables

Table 1: Demographic characteristics of study participants 20

Table 2: Clinical Presentation 20

Table 3 Time to failure of ETV in DWM patients 22

Table 4: Chi-square test for Age, sex, and Presence of infection 24

List of Figures

Figure 1 Overall survival probability of Patients after ETV with no need of Shunting from January 1, 2021- May 2023 23

Figure 2 . Kaplan-Meier Survival Plot of Patients with infection vs. patients with no infection. 24

Acronym and abbreviations

- CPC---------Choroid Plexus Cauterization
- CPS--------- Cystoperitoneal shunt
- CSF---------Cerebrospinal Fluid
- DWC--------Dandy Walker Complex
- DWM-------Dandy Walker Malformation
- DWV--------Dandy Walker Variant
- DWS---------Dandy Walker Syndrome
- ETV---------Endoscopic Third ventriculostomy
- VPS---------Ventriculoperitoneal shunt

Abstract

Background: *Dandy-Walker complex (DWC) is a term coined to describe a continuum of posterior fossa anomalies categorizing spectrum of cystic malformation posterior fossa*

depending on extent of involvement and severity. DWS has been reported to occur in 1 of 25,000 to 30,000 newborns, with the majority of patients presenting in the first year most of them present with hydrocephalus (70–90%). There are different treatment modalities to treat hydrocephalus in Dandy-walker Malformation patients endoscopic Third ventriculostomy being one of them. In our country Ethiopia, ETV has also been applicable for the treatment of the hydrocephalus in those patients with DWM lately. Despite its applicability in the management of Hydrocephalus its outcome hasn't been properly assessed. This research aims at assessing the treatment outcomes of ETV.

***Objectives**: To assess the treatment outcomes of Hydrocephalus among patients with DWM who are treated with ETV at Zewditu Memorial Hospital.*

***Methods**: Retrospectively collected data in 33 children with confirmed Dandy-Walker Malformation patients who have undergone ETV was conducted at Zewditu Memorial Hospital from January 2021 to May 2023. The Data collected was analyzed using SPSS. Frequency distribution will be used to describe the characteristics of the patients. Data analysis was performed using chi-squared tests and survival analysis by Kaplan-Meier tests to assess the treatment outcomes and survival probability.*

***Result:** The median age at surgery was 7 months and 84.7% of the patients were less than 1 year. There was slight Male predominance than females 1.75:1. ETV was successfully done in 22(66.7%) of the patients with improvement in their initial presenting clinical sign & symptoms. The 6-month ETV failure rate was 33.3% to whom we did redo ETV/VP-shunting. All failures occurred within the first 6 months after surgery. The estimated cumulative survival probability was 75.8% at 2.4 months, 70 % at 4 months and with mean follow up time of 18 months.*

***Conclusion:** The 6 month ETV failure rate was 33.3%. Endoscopic third ventriculostomy should be strongly considered as primary management in treating DWC associated hydrocephalus.*

***Key terms**: Dandy walker Malformation, Endoscopic third ventriculostomy, Hydrocephalus*

1. Introduction

1.1 Background

Dandy-Walker malformation (DWM) represents a congenital malformation characterized by agenesis or hypoplasia of the cerebellar vermis, cystic dilation of the fourth ventricle, and enlargement of the posterior fossa with or without hydrocephalus. It has been described with different imaging characteristics starting from coining of the name into a different timeline. (1)

The embryologic background of the DWM has been postulated by different scholar in various ways to explain the origin of this congenital malformation: the CSF production and dynamics leading to over production resulting in dilatation of fourth ventricle and later hypoplasia of surrounding structures; later has been given the theory of the persistence of the posterior medullary velum, which remains as a thick arachnoid and ectodermal membrane leading to fourth ventricular cystic dilatation and some other imply to, though not commonly seen, developmental failure of the foramen of Magendie. But in later studies it has been given to consider the pathogenesis DWM from the embryologic basis can be attributed to dysgenetic development of the anterior membranous area of the rhombencephalon. (2)

Dandy-Walker complex (DWC) is another term coined to describe a continuum of posterior fossa anomalies categorizing spectrum of cystic malformation posterior fossa depending on extent of involvement and severity. This gives a wider differential diagnosis for DWM and includes more cystic congenital malformations of posterior fossa. From this, Dandy walker Variant (DWV), Persistent Blake's pouch cyst, retro cerebellar arachnoid cyst and mega cisterna magna.(1)

Dandy walker syndrome (DWS) has been reported to occur in 1 of 25,000 to 30,000 newborns, with the majority of patients presenting in the first year in most of whom hydrocephalus develops in 70–90% of cases. Dandy Walker malformation is present in 2–4% of all cases of hydrocephalus. Clinical presentation tends to be somewhat age specific and the symptoms in DWM are related to hydrocephalus, cerebellar, and cranial nerves dysfunction and to the presence of associated anomalies.(1, 2)

Different treatment strategies have been put into action for the management of DWS-related hydrocephalus (DWSH), including a ventriculoperitoneal shunt (VPS), a cystoperitoneal shunt

(CPS), and endoscopic third ventriculostomy (ETV) with or without choroid plexus cauterization (CPC). (3, 4)

1.2 Statement of the Problem

Up to date the ideal treatment of DWS is not well stated and for decades, CSF diversion has been used as a main stay of treatment. It can be diverting the CSF from the lateral ventricles (supratentorial compartment) or the posterior fossa cyst (infratentorial compartment). Even in early days, cyst excision has been used as a treatment modality for DWS, which had higher morbidity and mortality of up to 10%. (5)

Diverting CSF with shunting has its own ease in treating HCP with simple Access to ventricle and also diverting the cyst with added risks of fatal complications. In doing shunts, the patient will be committed to lifelong shunt dependence and to multiple surgical correction, procedures that come with shunt malfunction and complications. (5)

On the other hand, endoscopic procedures, Endoscopic Third Ventriculostomy (ETV) can give end result outcome for hydrocephalus treatment in improving neurocognitive outcome of these babies. Apart from alleviating the shunt dependence and shunt related complications, stabilizes the ventricular size and this mode of treatment can be good option of management in treating DWS. (6)

In our country Ethiopia, ETV has also been applicable for the treatment of the hydrocephalus in those patients with DWM lately. Despite its applicability in the management of Hydrocephalus its outcome hasn't been properly assessed. This research aims at assessing the treatment outcomes of ETV filling in this gap.

1.3 Justification

DWS is one of the causes of congenital hydrocephalus in infants. And our main stay of treatment is addressing hydrocephalus with CSF diversion, with shunting of the ventricles or cyst, to achieve a good neurocognitive development and control of ventricular size in those infants. So, the use of ETV is coming to a picture in treating hydrocephalus and achieving the goals of treatment that has been attained with shunting, even with some added benefits in shunt free survival.

Despite the fact that few research has been done worldwide in comparing and assessing the factors that determine the outcome in treating DWS with shunting and ETV, there is no given and settled way of management of this congenital malformation.

Thus, this study will assess the outcomes of DWM patients being treated with ETV and assess the complications which are associated with ETV. This research will add more input in developing the best treatment option in managing DWM in our setup.

2. Literature Review

Endoscopic third ventriculostomy (ETV) can be an effective and appealing treatment for hydrocephalus; however, the procedure is not always successful. Varied success rates based on patient age, etiology of hydrocephalus, and prior shunting highlight the importance of patient selection for ETV.(7)

According to B. C. Warf and his colleagues a study done on Management of Dandy-Walker complex–associated infant hydrocephalus by combined endoscopic third ventriculostomy and choroid plexus cauterization the ETV/CPC (ETV alone in 1 patient) successfully treated the hydrocephalus in 31 patients(74%) among the 45 patients the study was done on with mean and median follow-up periods of 24.2 and 20 months, respectively, (range 6–65 months), whereas 10 (24%) required another operation for hydrocephalus. The study also has shown that the median age at ETV/CPC for those in whom treatment failed and for those in whom it was successful was 3.5 and 5 months, respectively. Two of the patients with DWM in whom treatment failed underwent endoscopic reopening of a closed ETV at 1.5 and 4 months, respectively. Both procedures were successful with no further operations at 34 and 9 months of follow-up. The probability of survival with no need for additional surgery was 69% at 1 year (95% CI 49%–88%) and 64% at 2 years (95% CI 38%–90%). The study has also shown that most of the treatment failures occurred prior to 6 months.(6)

Mohanty and Co-authors on their research on treatment options for Dandy-Walker Malformation also discussed the treatment outcomes of ETV and Shunt placement in treating Dandy walker Malformation cases. The study was done on 72 patients with DWM among which 21 patients had undergone ETV. Among 21 children who underwent ETV 5 of them developed CSF Collection for which CSF drainage through the patent anterior fontanel was done. Endoscopic procedures failed in six patients (24%), five of whom underwent placement of a VP shunt.(8)

Garg et al. on his research about ETV has shown that 99 patients from 115 patients (86.1%) improved, and the rest either worsened or continued to have the symptoms. The study also implied that most frequent complications seen after ETV were CSF leakage in 7 patients (6.1%), meningitis in 3 patients (2.6 %) and seizure in 2 (1.7%) patients, which were managed successfully with conservative treatment.(9)

A study done on treatment strategies for hydrocephalus related to Dandy-Walker syndrome the 6-month ETV failure rate was 36.7%, and the overall failure rate was 51% which was comparable with fifty percent failure rate of the shunt-based treatment. According to the study patients in whom ETV-based treatment failed were younger (mean age at treatment 13.8 vs 24 months, $p = 0.037$). (10)

3. Objectives

3.1 General Objectives

- To assess the treatment outcomes and complications of ETV among patients with DWM treated for Hydrocephalus at Zewditu Memorial Hospital from January 2021-May 2023.

3.2 Specific Objectives

- To assess the treatment outcomes of Hydrocephalus among patients with DWM who are treated with ETV at Zeweditu Memorial Hospital from January 01, 2021, to May 01, 2023.
- To assess complications of ETV among patients with DWM with Hydrocephalus who are treated with ETV at Zewditu Memorial Hospital from January 01, 2021, to May 01, 2023.

4. Methods and materials

4.1 Study Area

Zewditu Memorial Hospital is a hospital in central Addis Ababa, Ethiopia. The hospital is named after Empress Zewditu . Today, Zewditu Memorial Hospital is operated by the Ministry of Health.

The neurosurgical unit at Zewditu memorial hospital is an affiliate department from Black lion hospital mainly managing pediatrics neurosurgery cases. Additionally, Neurotrauma and other neurosurgical pathologies are being managed at the hospital. The neurosurgical unit has a total of 35 beds of which 28beds are for pediatric patients and the remaining 7beds are for adult neurosurgical cases. The Out-patient department sees in total of 4000 pediatric cases per year (this number includes both new patient and those who have continuous follow up at the hospital). Around 290 hydrocephalus cases are operated per year at the hospital.

4.2 Study period

From January 01, 2021, to May 01, 2023

4.3 Study design

Retrospective Chart review

4.4 Source and study Population

4.4.1 Source Population:

- All Patients with congenital hydrocephalus who had confirmed DWM with CT-scan/MRI visiting Zewditu Memorial Hospital OPD clinic during the study period.

4.4.2 Study Population:

- All Patients with congenital hydrocephalus who had confirmed DWM with CT-scan/MRI visiting Zewditu Memorial Hospital OPD clinic during the study period who fulfill the inclusion criteria.

4.5. Eligibility criteria

4.5.1 Inclusion criteria

Patients who had congenital hydrocephalus and confirmed DWM with CT-scan/MRI imaging

4.5.2 Exclusion Criteria

- Patients who had congenital hydrocephalus having DWM with CT scan findings unfavorable for ETV (overcrowded prepontine space, signs of Scaring, those patients with concomitant post-infectious changes)
- Patents who were unable to provide adequate information and whose charts are incomplete.

4.6 Sample Size

All patients who were admitted for the diagnosis of Dandy walker Complex and treated with ETV as a primary treatment were taken as sample.

4.7Operational Definition

Dandy Walker Complex: is a new term to represent a continuum of different posterior fossa fourth ventricular cyst lesions including Dandy walker malformation, the dandy walker variant and mega cisterna magna.(11)

Dandy Walker malformation: an enlarged posterior fossa with partial (hypoplasia) or complete agenesis of the cerebellar vermis and cystic dilatation of the fourth ventricle which is distorted and encased in a neuroglial vascular membrane. (1)

Dandy Walker variant: when not all of the Dandy Walker malformation criteria are present. E.g., vermian hypoplasia and cystic dilatation of the 4th ventricle, without enlargement of the posterior fossa. (1)

Mega cisterna magna: enlarged posterior fossa secondary to an enlarged cisterna magna. Normal vermis and fourth ventricle and no mass effect on the cerebellum.(1)

ETV: Endoscopic third ventriculostomy is a procedure done to divert the CSF by opening third ventricular floor to communicate the ventricular system with the subarachnoid space.(2)

ETV Success: amelioration of the signs and symptoms of elevated intracranial pressure (ICP) in patients previously showing signs of Hydrocephalus and free from requiring subsequent surgery or death related to hydrocephalus management within six months of operation. (2, 7)

ETV Failure: cases requiring any subsequent surgical procedure for cerebrospinal fluid diversion after the primary management or case mortality below six months post-surgery.(7)

4.9 Study Variable

Dependent variable

- Treatment Outcome (Hydrocephalus Improvement measured by Head circumference and improvement in clinical manifestations during subsequent follow up Starting from 2 weeks)

Independent Variable

- Age at time of surgery
- CT-scan Findings favorable for ETV
- HC at presentation
- Sign of infection

4.10 Data Collection tool procedure

Data was collected by trained one General Practitioner who work at the Hospital by using structured questioner and physical examination during follow up visits starting from two weeks post ETV followed by every month visit for the first 6 months post ETV then every 6 month till the end of the study period. Patients were followed for a minimum of 6 months after surgery to be considered as successful ETV. Each patient's medical chart was coded with specific designation to avoid repetition in recording. Hydrocephalus was measured using the tape meter in occiputo-frontal manner each time they come for routine medical visit.

4.11 Data Quality Assurance

Data was collected using an interviewer administered questionnaire after training was provided to the data collectors on the details of the questionnaire. One General Practitioner and one supervisor participated in the data collection process. The data collectors were given one day training on the objectives of the study and different sections of the questionnaire, and interviewing techniques by the researcher.

Questionnaire filled were checked for its completeness and consistency daily by the assigned supervisor. The investigator was responsible for monitoring the overall data collection process and providing supportive supervision on the spot.

The data collected was carefully entered and cleaned before analysis.

4.12 Data Processing and analysis

The collected data was coded and recorded into an excel spread sheet and checked for consistency and completeness before being exported to computer Software for Statistical

Package for Social Sciences (SPSS) version 25 for the required analysis. Descriptive statistics such as frequencies mean median and standard deviation were used to summarize participants' characteristics. Chi-squared analysis was performed using the Fisher 2-tailed test that was used to test the significance of differences among groups, with $p < 0.05$ being considered significant. Survival and treatment success probabilities were calculated using the Kaplan-Meier method.

4.13 Ethical Consideration

Before data collection to conduct this study, ethical clearance was obtained from the institutional review board (IRB) at Black Lion Specialized Hospital and submitted to Zewditu Memorial hospital research affairs. The aim of the study was clearly explained to the study participants and their right to refuse was maintained. Information was collected after obtaining informed verbal consent from each participant's care givers. The personal information of study participants was kept entirely anonymous, and confidentiality was assured throughout the study period. The name and address of the patient was omitted from the questioner.

4.14 Dissemination of results

After being completed, the research paper was submitted to Black Lion Specialized Hospital Neurosurgery department. The findings of this study will be distributed to Black Lion Specialized Hospital, Zewditu Memorial Hospital. If possible, the findings will be presented in different seminars, meetings and workshops and will be published in scientific journals.

5. Results

Demographic characteristics

Among the 33 DWC participants in our study the majority were male 21(63.6%). At the time of treatment patients had mean and median age of 11.39 months (SD 13.5) and 7 months, respectively. From the 33 DWC patients 39.4% of them were younger than 6 months at the time of treatment, 84.8% younger than 12 months old and 15% of them were older than two years old. Our study has shown slight male predominance over female participants (ratio 1.75:1) (Table 1)

Table 1: Demographic characteristics of study participants

		ETV (n=33)
Age n (%)	1-6 month	13 (39.3%)
	7-12 month	15 (45.4%
	>12 month	5 (15.1%)
Sex n (%)	Male	21 (63.6%)
	Female	12 (36.4%)

Clinical presentation

All of our study participants presented with increased in head circumference. Up-gaze palsy being the second most common presenting symptom (33.3%) followed by Irritability and bulged fontanel (18.2%). Less than 5% of our study participants presented with seizure and extremity weakness. (Table 2)

Table 2: Clinical Presentation

Symptoms	**ETV n=33**
Vomiting	9.1%
Irritability	18.2%
Seizure	3%
Poor feeding	6.1%
Physical finding	
Increased HC	100%
Bulged fontanelle	18.2%
Up gaze palsy	33.3%
Extremity weakness	3%

Neuro-imaging Evaluation

All of our patients were imaged preoperatively with CT/MRI after thorough clinical evaluation, when they come to our outpatient clinic. Most of these patients had CT scan and three from the total patients had MRI. All the imaging findings were critically evaluated with the team of neurosurgical consultants, and some are also commented with consultant neuro-radiologists. This was done for the delicacy of the issue in differentiating the diseases spectrum from those similar imaging findings in other pathologies. We eliminated those patients with enlarged posterior fossa cyst consistent with posterior fossa Arachnoid cyst. We used our patients preoperative CT to put our patients as DWM and DWV as per the definitions established in literatures. (11) We defined DWM as: 1) complete absence of the vermis, or hypoplasia of the inferior vermis; 2) a posterior fossa cystic CSF collection in direct communication with the fourth ventricle; and 3) an enlarged posterior fossa with elevation of tentorium and torcula herophili. And DWV was defined as: 1) inferior vermis hypoplasia; 2) a posterior fossa CSF collection with direct communication to the fourth ventricle; and 3) no obvious enlargement of the posterior fossa was seen. (11) Depending on the above definition we had four patients with DWV. We had three patients with MRI which one of them has imaging features of MCM and one with persistent BPC. All of the patients were having ventriculomegaly which their common presentation was also with enlarged head size and imaging evaluation of the Aqueduct was open in all. To obtain post-operative imaging in our patients were so difficult because of the financial constraint the parents have to afford for the imaging.

Complications

Among the 33 participants who underwent ETV almost all were accomplished with minimal or no bleeding except one patient who had intra-OP haemorrhage that was controlled immediately. Post-operative CSF leak was found in 12(36.4%) our participants for whom we did wound reinforcement by applying interrupted sutures with application of pressure dressing, and 7(21.2%) of the patients developed infection (ventriculitis, wound site infection).

Treatment Outcome

From the 33 DWS patients, 7(21%) had undergone ETV/CPC with greater than 90% Cauterization of the choroid plexus and 2(6%) of our patients underwent ETV/cyst fenestration of posterior fossa and the remaining 24(73%) underwent ETV alone. ETV was successfully done

in 22(66.7%) of the patients with improvement in their initial presenting clinical sign & symptoms. We had 4 patients who are lost to follow up and one patient died of unknown cause at home 4 months after surgery.

The 6-month ETV failure rate was 33.3% to whom we did redo ETV/VP-shunting. All failures occurred within the first 6 months after surgery. *(Table 3)* Most of (80%) the patients who had ETV failure happen to be younger than 6 months at the time of surgery. The reasons for failure were not recorded in all of the medical charts.

Table 3 Time to failure of ETV in DWM patients

Time to failure	No of failed ETV	Per cent
< 1 month	**1**	**3%**
1-6months	**10**	**30.3%**
> 6months	**0**	**0**
Total no of ETV failure	**11**	**33.3%**

In 26 months of follow up, the probability of survival with no need for shunting or redo was 66.2% at 6 months, 1 year and 2 years. The estimated cumulative survival probability was 75.8% at 2.4 months, 70 % at 4 months with mean follow up time of 18 months (95% CI, 14.32-22.14). The median survival probability was not estimated in this study given the short duration of the study (<3 years) implying the need to follow for more months or years.

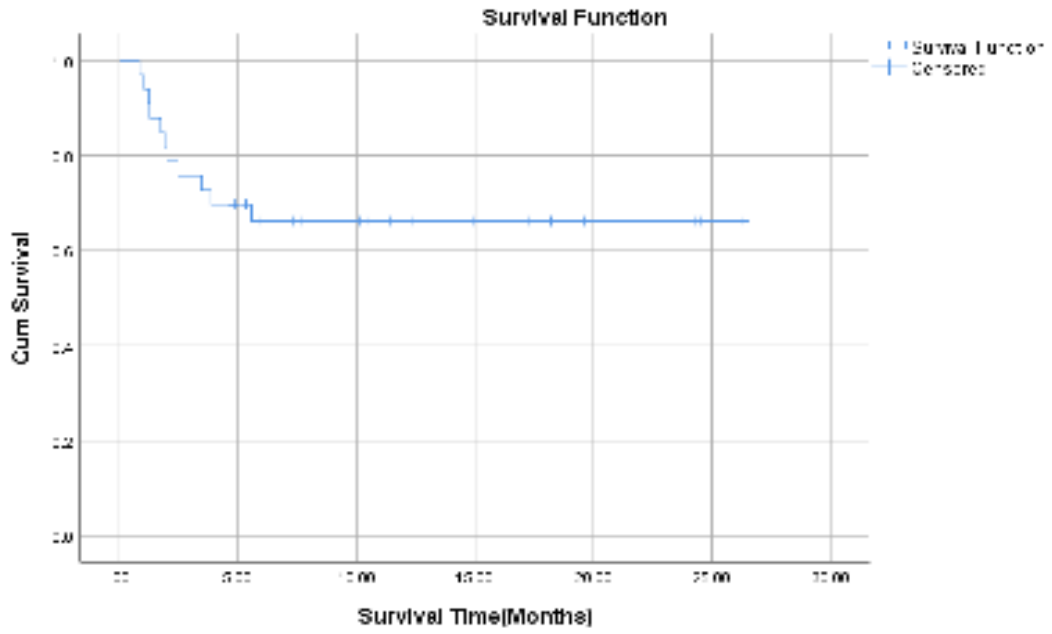

Figure 1 Overall survival probability of Patients after ETV with no need of Shunting from January 1, 2021- May 2023

Log-rank test was performed to test if there were any significant variation in survival times among groups. In accordance to the test those with infection appear to have failure earlier than those who with no infection (log-rank test, p=0.002)

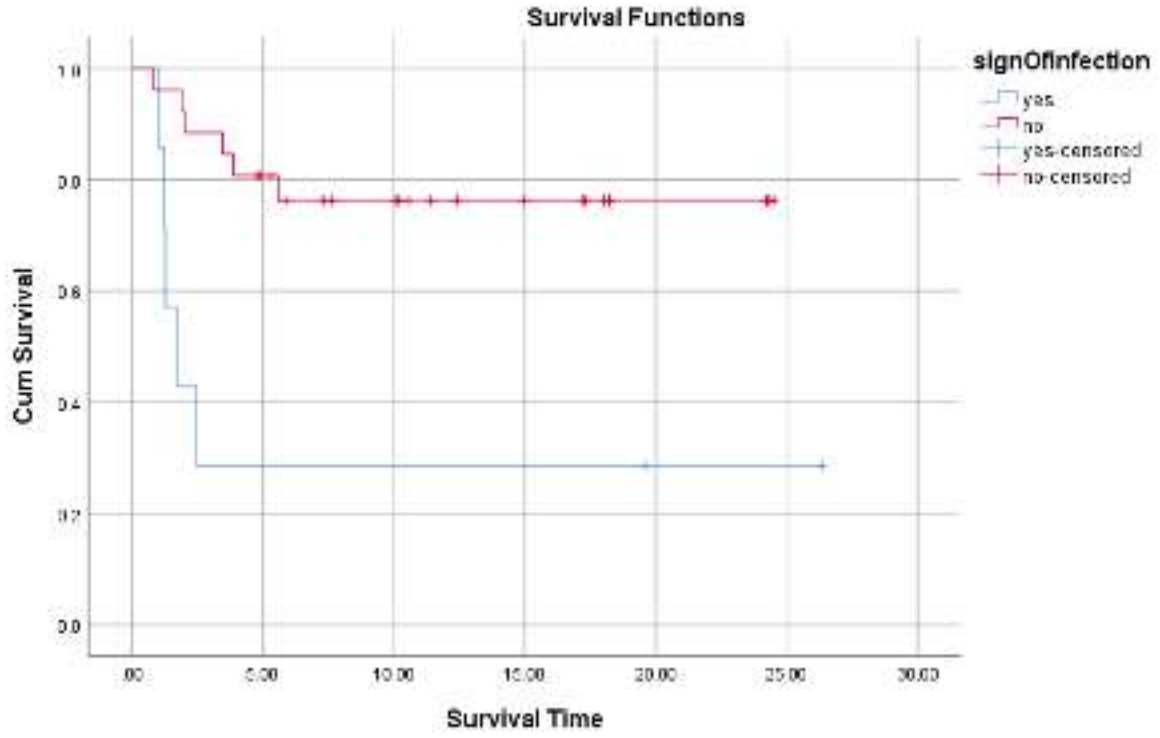

Figure 2 . Kaplan-Meier Survival Plot of Patients with infection vs. patients with no infection.

Using Fischer's exact test our study showed significant association between age (age < 6months vs. age ≥ 24 months, p=0.003) and presence of infection (p= 0.027) with ETV outcome. No significant differences were seen between other patient characteristics and ETV outcome. (Table 4)

Table 4: Fischer's test for Age, sex, and Presence of infection

Variables		**ETV Outcome**		**Value**	**p-value**
		Success	**Failure**		
Age	**1-6 month**	4	9	**11.6**	**0.03***
	7-12 month	13	2		
	>12 month	5	0		
Sex	**Male**	13	8	**0.589**	**0.703**

	Female	9	3		
Infection	**Yes**	20	2	**5.802**	**0.027***
	No	6	5		
CSF leak	**Yes**	5	7		**0.052**
	No	17	4		

In cox-regression analysis was done for age, Presence of CSF leak, and presence of infection. The result revealed that only patients with infection have shown higher failure (HR=5.230, 95% CI 1.316-20.779, p=0.019). Otherwise, Age (HR=0.756, 95% CI 0.581-0.983, p=0.037) and those with CSF leak (HR3.240, 95% CI 0.839-12.519, p= 0.088) have shown no significant association.

6. Discussion

With the growing acceptance of neuro-endoscopy, the traditional way of managing obstructive hydrocephalus by shunting is now less favoured due to shunt dependency and malfunction.

In this study I tried to assess the treatment outcome of ETV in those patients with DWM starting from January 2019 to May 2023. ETV based primary treatment of hydrocephalus in those patients with DWM has been recently implemented in our country and its efficacy compared to shunt-based management has not been properly assessed. To my knowledge this research is the first to be conducted in our country assessing the treatment outcomes of ETV in those patients with DWM.

ETV has been reported successful in managing obstructive hydrocephalus due to Dandy walker Complex in 49-94% of cases.(9, 12) In this study the success rate of ETV in treating hydrocephalus with no need for shunting is 66.7%. This report is slightly lower than the success rates reported in different studies. According to Warf BC et al a study conducted in Uganda 74% success rate was reported this difference in success rate might be due to the higher rates of ETV/ CPC conducted in their study as compared to ours.(6) Two out of the six patients who had ETV/CPC in our case had undergone shunting while the remaining 4 had shown improvement clinically post-surgery. Another study done in 15 patients with DWM by Garg and Co-authors reported 93.3% success rate of ETV combined with aqueductoplasty and cystoventriculostomy. (9)

This study showed that all ETV failures occurred in the first 6 months post-surgery which is consistent with studies.(8, 10) Although most ETV failures occur in the first 6 months after surgery Warf has reported 1 patient that who had failure 1 year after ETV suggesting the possibility of late failures.(6)

The probability of survival at 6 months, 1 year and 2 years is similar (66.2%) in this study. This survival probability is comparable to the study done by Warf and his colleagues showing 69%, 64% survival probability at 1 and 2 years respectively with mean follow up time of 24 months.(6) A study done in North- America by Aaron et al has shown 49 % survival probability and ETV success in those patients treated with ETV which is relatively lower than survival probability reported in our study. (10)

Most of the patients in this study (84.7%) are less than 1 year and 39.3% are below 6 months old. The median age at the time of treatment was 7 months. This is in contrast to Mohanty and co-authors where 50% of the patients were younger than 1 year and 31% of them were above 2 years.(8) The higher number of younger patients in this study might explain the lower success rate of ETV as compared to some studies since younger children have lower ETV success score (8, 13).

Similar to other studies CSF leak was the commonest post-op complication reported in this study (8, 10). Additionally, patients with infection had higher failure rates as compared to those with no infection. This might be due to an early closure/obstruction of the created stoma in the floor of third ventricle by the inflammatory reaction induced by the infection, even sometimes extensive enough to form secondary Arachnoid layer, dense fibrin accumulation or scaring in the cisternal space.(14, 15)

7. Strength and Limitations

7.1 Strength

To my knowledge this the first research conducted in Ethiopia assessing the treatment outcome of ETV in DWC patients. We have tried to assess the Success rate of ETV along with its complication in detailed manner.

7.2 Limitation

Our Study only assessed those patients who are confirmed DWC patients via CT-Scan or MRI imaging. Those patients who could not afford CT- Scan or MRI for diagnosis of DWC underwent Shunting without knowing the actual cause of hydrocephalus. Therefore, we might have missed few DWC patients who could not afford to pay for imaging modalities. This study only assessed the treatment outcome of ETV in DWM patients. Comparing the success rate of Shunting and ETV in DWC patients might give us more information in choosing the better primary intervention for the treatment of DWC associated hydrocephalus. Our study is conducted only in one Hospital and multi-centre approach will increase our sample size and generalizability.

8. Conclusion and Recommendation

The role of shunts in the treatment of congenital hydrocephalus in patients with DWM is already well established but there is always a concern of shunt dependence and malfunction. These complications and living in third world countries are additional burden to patients as well as family members. With the growing acceptance of ETV, shunting is now becoming less favoured. In this study we have reported 66.6% of survival probability with no need for shunting with mean follow up duration of 18 months. CSF leak and infection (ventriculitis and wound site infection) being commonest complications. Presence of infection after surgery seems to be predictor for ETV failure. Interventions to reduce the chance of developing infection should be implemented to reduce ETV failure. In general, ETV should be strongly considered as primary management in treating DWC associated hydrocephalus.

9. Reference

1. Greenberg MS. Greenberg's Handbook of Neurosurgery.10th Edition:270-73.

2. H. Richard Winn M. Youmans & Winn Neurological Surgery.7th Edition:5843-51.

3. Brusius CV, Cavalheiro S. Endoscopic third ventriculostomy is a safe and effective procedure for the treatment of Blake's pouch cyst. Arq Neuropsiquiatr. 2013;71(8):545-8.

4. Hu CF, Fan HC, Chang CF, Wang CC, Chen SJ. Successful treatment of Dandy-Walker syndrome by endoscopic third ventriculostomy in a 6-month-old girl with progressive hydrocephalus: a case report and literature review. Pediatr Neonatol. 2011;52(1):42-5.

5. Kulkarni AV, Riva-Cambrin J, Browd SR, Drake JM, Holubkov R, Kestle JR, et al. Endoscopic third ventriculostomy and choroid plexus cauterization in infants with hydrocephalus: a retrospective Hydrocephalus Clinical Research Network study. J Neurosurg Pediatr. 2014;14(3):224-9.

6. Warf BC, Dewan M, Mugamba J. Management of Dandy-Walker complex-associated infant hydrocephalus by combined endoscopic third ventriculostomy and choroid plexus cauterization. J Neurosurg Pediatrics. 2011;8(4):377-83.

7. Deopujari CE, Karmarkar VS, Shaikh ST. Endoscopic Third Ventriculostomy: Success and Failure. J Korean Neurosurg Soc. 2017;60(3):306-14.

8. AARONMOHANTY MC, ARUNDHATIBISWAS, M.CH., SATYANARAYANASATISH, M.CH.,, SHANTISHANKARPRAHARAJ MC, ANDKOLLURIV. R. SASTRY, M.CH. Treatment options for Dandy–Walker malformation. J Neurosurg: Pediatrics November, 2006;Volume 105.

9. Garg A, Suri A, Chandra PS, Kumar R, Sharma BS, Mahapatra AK. Endoscopic third ventriculostomy: 5 years' experience at the All India Institute of Medical Sciences. Pediatr Neurosurg. 2009;45(1):1-5.

10. Yengo-Kahn AM, Wellons JC, Hankinson TC, Hauptman JS, Jackson EM, Jensen H, et al. Treatment strategies for hydrocephalus related to Dandy-Walker syndrome: evaluating procedure selection and success within the Hydrocephalus Clinical Research Network. J Neurosurg Pediatr. 2021;28(1):93-101.

11. Barkovich AJ, Kjos B, Norman D, S.Edwards M. Revised Classification of Posterior Fossa Cysts and Cystlike Malformations Based on the Results of Multiplanar MR Imaging. AJNR 10:. December 1989.

12. Zandian A, Haffner M, Johnson J, Rozzelle CJ, Tubbs RS, Loukas M. Endoscopic third ventriculostomy with/without choroid plexus cauterization for hydrocephalus due to hemorrhage, infection, Dandy-Walker malformation, and neural tube defect: a meta-analysis. Childs Nerv Syst. 2014;30(4):571-8.

13. Kulkarni AV, Riva-Cambrin J, Holubkov R, Browd SR, Cochrane DD, Drake JM, et al. Endoscopic third ventriculostomy in children: prospective, multicenter results from the Hydrocephalus Clinical Research Network. J Neurosurg Pediatr. 2016;18(4):423-9.

14. WOLFGANGWAGNER A. Failure after endoscopic third ventriculostomy. J Neurosurg (Pediatrics). Juluy 2005;vol 103.

15. Moreira I, Pereira J, Oliveira J, Salvador SF, Vaz R. Endoscopic re-opening of third ventriculostomy: Case series and review of literature. Clin Neurol Neurosurg. 2016;145:58-63.

Annex

Questionnaire Tool

AAU

COLLEGE OF MEDICINE AND HEALTH SCIENCE

Department of Surgery Neurosurgery Unit

This study is to be done by Neurosurgery resident at Black Lion Hospital. The purpose of this data collection tool is to capture data regarding Treatment Outcome of Hydrocephalus among patients with Dandy Walker Malformation managed with ETV at Zewditu Memorial Hospital, from January 2021- June 2023. This structured data collection tool will use data collected routinely at Zewditu memorial hospital, data entered on patient chart and direct interview with the attendant of the children for the treatment outcome.

It will take 15 to 20 minutes for the data collection process. This study does not bring any harm to your health. If you feel discomfort with the questions and the process of measurement, please feel free to drop at any time you want. Please be assured that your confidentiality is well preserved, and also you have the right not to participate in the study, but your honest participation is important.

The tool has three sections assessing socio-demographic, clinical feature, Imaging finding, Intraoperative finding, Treatment outcome at each follow up and developmental assessment tool. It is very important to ensure the quality of data collected since it will be used to plan appropriate measures that could be taken.

Unique ID- _____

Date of data collection-_____

Name of data collector _____

Name of supervisor-_____

Data collection completeness: Not completed ☐ Completed ☐ Partially completed ☐

Section 1: Socio-demographic status	
Residency	1.Urban 2. Rural
Sex	1. Male 2. Female
Section 2: Clinical Presentation	
Hydrocephalus	Head circumference initially _____cm
Neurological examination	Presence of extremity weakness_____ Level of consciousness - A. Alert - B. Lethargic - C. Comatose
Seizure	1.Yes 2. No
Vomiting	1. Yes 2. No
Anterior Fontanel Status	1.Soft 2.Bulged and Tense
Poor feeding	1.Yes 2.No
Section 3: CT- Scan initial finding	

Section 4: Intraoperative Finding (Presence of bleeding, Presence of prepontine scarring, degree of cauterization if CPC done)

Section 5: Treatment Outcome							
Date ETV was done_____ Age ETV was Done_____							
How many weeks/Months since the ETV_____							
Post ETV Follow up							
	2nd week	1 month	2nd month	3rd month	4th month	5th month	6th month
1. Improvement of presenting Symptoms							
Decrement in Head Circumference	1.Yes 2.No	1. Yes 2.No	1.Yes 2.No	1.Yes 2.No	1.Yes 2.No	1.Yes 2.No	1.Yes 2.No
Neurological deficit improvement	1.Yes 2.No	1.Yes 2.No	1.Yes 2.No	1.Yes 2.No	1.Yes 2.No	1.Yes 2.No	1.Yes 2.No
Level of consciousness improvement	1.Yes 2.No	1.Yes 2.No	1.Yes 2.No	1.Yes 2.No	1.Yes 2.No	1.Yes 2.No	1.Yes 2.No
Seizure	1.Yes 2.No	1.Yes 2.No	1.Yes 2.No	1.Yes 2.No	1.Yes 2.No	1.Yes 2.No	1.Yes 2.No

Vomiting	1.Yes 2.No	1.Yes 2.No	1.Yes 2.No	1.Yes 2.No	1.Yes 2.No	1.Yes 2.No	1.Yes 2.No
2. Patient needs redo ETV or shunt placement	1.Yes 2.No	1.Yes 2.No	1.Yes 2.No	1.Yes 2.No	1.Yes 2.No	1.Yes 2.No	1.Yes 2.No
3. Patient died of causes related to Hydrocephalus	1.Yes 2.No	1.Yes 2.No	1.Yes 2.No	1.Yes 2.No	1.Yes 2.No	1.Yes 2.No	1.Yes 2.No
4. Presence of CSF Leak	1.Yes 2.No	1.Yes 2.No	1.Yes 2.No	1.Yes 2.No	1.Yes 2.No	1.Yes 2.No	1.Yes 2.No
5. Signs of Infection							
5.1 CBC elevated.	1.Yes 2.No	1.Yes 2.No	1.Yes 2.No	1.Yes 2.No	1.Yes 2.No	1.Yes 2.No	1.Yes 2.No
5.2 CSF Analysis Suggestive	1.Yes 2.No	1.Yes 2.No	1.Yes 2.No	1.Yes 2.No	1.Yes 2.No	1.Yes 2.No	1.Yes 2.No
5.3 Fever	1.Yes 2.No	1.Yes 2.No	1.Yes 2.No	1.Yes 2.No	1.Yes 2.No	1.Yes 2.No	1.Yes 2.No
6.Intraopertaive hemorrhage	1.Yes 2.No	1.Yes 2.No	1.Yes 2.No	1.Yes 2.No	1.Yes 2.No	1.Yes 2.No	1.Yes 2.No
7. Wound site infection	1.Yes 2.No	1.Yes 2.No	1.Yes 2.No	1.Yes 2.No	1.Yes 2.No	1.Yes 2.No	1.Yes 2.No

APPROVAL SHEET

Addis Ababa University,

School of Medicine, Department of surgery, Neurosurgery Unit

I hereby, certify that I have read and evaluated this topic entitled Treatment Outcome of Hydrocephalus among patients with Dandy Walker Malformation managed with ETV at Zewditu Memorial Hospital, from January 2021- May 2023, Addis Ababa, Ethiopia, which was prepared under my guidance. I recommend it to be submitted as fulfilling the thesis requirement.

Submitted by

- **Dr. Bereket Hailu _____ __ /__ /__**

  Full name Signature Date

Primary Advisors

- **Dr. Kibruyisfaw Zewdie _____ __ /__ /__**

  Full name Signature Date

- **Dr. Yemisirach Bizuneh _____ __ /__ /__**

  Full name Signature Date