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## Original Article

# Endoscopic Third Ventriculostomy in the Treatment of Late-Onset Idiopathic Aqueductal Stenosis: Outcome Evaluation

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**BACKGROUND:** Endoscopic third ventriculostomy is the shunt-independent surgical treatment for hydrocephalus caused by late-onset idiopathic aqueductal stenosis.

**OBJECTIVE:** This study aims to evaluate the success rate and the clinical and radiological outcomes of endoscopic third ventriculostomy in late-onset idiopathic aqueductal stenosis.

**PATIENTS AND METHODS:** A retrospective analysis was conducted on 15 patients (9 males and 6 females) with hydrocephalus due to late-onset idiopathic aqueductal stenosis in whom endoscopic third ventriculostomy was performed. Different clinical, radiological and operative factors affecting the outcome were analyzed.

**RESULTS:** Twelve patients in our study had a chronic clinical history (class III), and 3 cases had subacute symptoms (class II). Success was obtained in 14 patients (93.3%) while failure was reported in one patient only (6.7%). Headache and gait disturbance improved in all patients, while 5 of the 8 patients with cognitive impairment and 4 of the 6 patients with urinary incontinence showed improvement. The postoperative images showed radiological improvement in 14 patients, with evident reduction in ventricular size in only 3 patients (20%).

**CONCLUSION:** Endoscopic third ventriculostomy is a safe, effective, and highly successful procedure in the treatment of hydrocephalus due to idiopathic aqueductal stenosis, with more than 93.3% success rate. To enhance our findings, further carefully planned, prospective, multicentric studies on bigger samples should be performed.

**KEYWORDS:** Aqueductal stenosis, Endoscopic third ventriculostomy, Idiopathic, Late-onset, Triventricular hydrocephalus.

## INTRODUCTION

Stenosis of the cerebral aqueduct encountered in infancy is mostly attributed to intrauterine infection or hemorrhage and it presents early with obstructive hydrocephalus and head enlargement. On the other hand, aqueductal stenosis may present later in childhood or adults. The underlying pathology may be the result of either space-occupying lesions (Such as tectal gliomas, hamartomas, posterior fossa, or pineal tumors), or primitive stenosis. Late-onset idiopathic aqueductal stenosis (LIAS) is a primitive obstructive hydrocephalus presented in adulthood.[1,2] The pathophysiological mechanism of the disease was thought to be malformative or inflammatory, the latter is due to an aberrant proliferation of subependymal glia of the aqueduct of Sylvius.[3]

The clinical presentation in patients with late-onset aqueductal stenosis is greatly variable.[4,5] The symptoms of LIAS typically resemble those of normal pressure hydrocephalus (More than one of the Hakim symptom triad of cognitive decline, gait disturbances, and urine incontinence), though they can occasionally coexist with obstructive hydrocephalus symptoms brought on by an increase in intracranial pressure (ICP), such as headache, endocrinological/visual/ocular disturbances, extrapyramidal signs, etc.[1,6] The clinical classification of LIAS was first suggested by Fukuhara and Luciano. Furthermore, they divided patients in relation to disease duration into three categories; class I (Acute): symptoms lasting no more than one month, class II (Subacute): 1-6 months of symptoms, and class III (chronic): symptoms lasting more than six months.[6]

Endoscopic third ventriculostomy (ETV) is considered an effective, minimally invasive, shunt-independent surgical treatment of LIAS.[7] A successful ETV procedure means that neither shunt placement nor another endoscopic procedure was needed for further control of hydrocephalus in that patient by one year.[8] The success rate of ETV reported in the literature varies from 69 to 95%.[9-11] However, variable percentages of failed cases were reported in different studies.[12,13]

We assessed the effectiveness of ETV in treating patients with LIAS in our retrospective study. The clinical presentations, the preoperative and postoperative radiological findings, the operative findings and the clinical and radiological outcomes were analyzed.

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## PATIENTS AND METHODS

This retrospective study included 15 patients with hydrocephalus due to late-onset aqueductal stenosis, who were subjected to ETV at our Neurosurgery department from June 2016 to January 2021. Patients with hydrocephalus caused by any pathology other than idiopathic aqueductal stenosis and previously shunted patients were excluded. This study was approved by the Institutional Review Board (IRB): 34716. Before surgery, all patients signed an informed consent after a simple explanation of the benefits and possible complications of the surgical procedure.

Patient history including age, gender, clinical presentation, duration of symptoms, and previous traumatic, hemorrhagic, or infectious central nervous system (CNS) insults was meticulously revised. Preoperative magnetic resonance imaging (MRI) scans were re-evaluated to determine the degree of ventriculomegaly, sulcal effacement, transependymal permeation, third ventricular floor anatomy, and the pathological anatomy of aqueductal stenosis.

### Surgical Procedure

All patients were operated in the supine position under general anesthesia with their heads slightly elevated. A pre-coronal burr hole was performed through a small linear or C-shaped skin incision. Dura was coagulated and incised. The endoscope (Karl Storz SE & Co. KG, Tuttlingen, Germany) was introduced to the lateral ventricle and the foramen of Monro was identified to access the third ventricle. The site of ventriculostomy was determined based on the third ventricular floor anatomy. The ventriculostomy was performed using a blunt probe and was gradually dilated using a Fogarty catheter (Edward Lifesciences, Irvine, California, USA). The final size of the stoma should exceed 6-8 mm. The cortical track was plugged with Gelfoam (Baxter Healthcare Corporation, Hayward, California, USA) after hemostasis and the wound was closed in layers.

### Follow-up

Patients were discharged to the ward and followed up clinically for any postoperative sequelae like fever, vomiting, or fits. Patients were discharged on the next day and followed up for a minimum of six months. A postoperative computerized tomography (CT) scan was performed one week after surgery and MRI was performed one month later.

Various clinical, radiological and operative factors affecting the outcome were analyzed. The success of ETV was defined by clinical and radiological improvement with no shunt needed within one year of surgery.

### Statistical Analysis

Data was expressed as percentages calculated using a statistical package for social sciences statistical analysis software (IBM SPSS Statistics for Windows, IBM Corp, Version 22.0. Armonk, NY).

## RESULTS

This study included 15 patients (9 males and 6 females) whose ages ranged between 27 and 66 years with a median age of 51 years. Twelve patients presented with chronic clinical history (class III), and 3 cases presented with subacute manifestations (class II). All patients had progressive clinical manifestations related to impaired and/or decompensated cerebrospinal fluid (CSF) circulation due to obstruction of the aqueduct of Sylvius. The duration of symptoms before diagnosis ranged between 3-45 weeks, with a mean of 18 weeks. All patients had triventricular hydrocephalus of variable degrees due to aqueductal stenosis. Cortical sulci effacement was encountered in 13 patients (86.6%) and CSF permeation was detected in six patients (40%). Complete ETV was accomplished in all patients. One patient had septum pellucidum defect and a second Lilliquist's membrane was encountered in another patient. Two patients had membranous scarring, obliterating the foramen of Monro in one patient and the subarachnoid space of the prepontine cistern in the other. The patients' demographic characteristics, clinical presentations, radiological and operative findings and complications were summarized in **(Table 1).**

Patients were evaluated after an average follow-up period of 32 (21–52) months from surgery. The clinical outcome at the end of the follow-up period was summarized in Table 2. Five patients presented with headache at the time of surgery; headache was improved in 4 patients, with complete resolution of headache in the fifth patient. Five of the 12 patients with gait disturbance had complete resolution of their unsteady gait, and the remaining 7 patients showed partial improvement. Of the 8 patients who presented with cognitive decline, 2 patients resumed their prior social and professional lives, 3 patients showed improvement, and the other 3 patients had no change. Six patients had urinary incontinence, 4 of them exhibited improvement, while the other 2 had no change in their condition. The postoperative images showed radiological improvement in 14 patients but did not reach normal size **(Fig. 1).** However, evident ventricular size reduction was obtained in three patients only (20%).

Transient postoperative fever and/or vomiting were the most common postoperative sequelae found in four patients. Only one patient had transient diabetes insipidus postoperatively. Fourteen patients needed no further CSF diversion and were considered successful with a success rate of 93.3%. Only one patient underwent unsuccessful ETV; this patient had postoperative meningitis that was treated with intrathecal and intravenous antibiotic injections but later developed secondary communicating hydrocephalus and required a ventriculoperitoneal (VP) shunt.

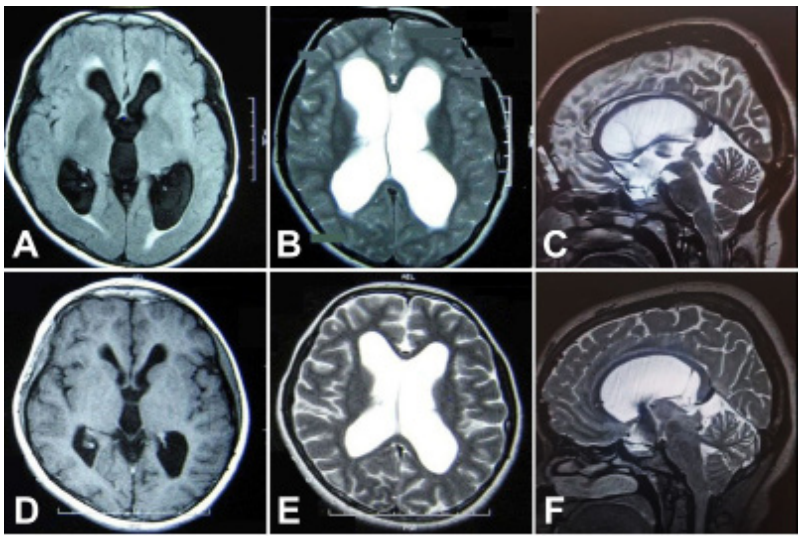


**Fig 1: (A, B, C) Preoperative MRI (axial flair, axial T2-weighted image and saggital T2-weighted image) of a case of LIAS showing evident supratentorial ventriculomegaly with CSF permeation and cortical sulci effacement, associated with downward bulging of the third ventricle floor and CSF pathway obstruction at the aqueduct. (D, E, F) Postoperative MRI (axial T1-weighted image, axial T2-weighted image and saggital T2-weighted image) showing successful communication between the third ventricle and the prepontine cistern.**

**Table 1: Patients' demographic characteristics, clinical presentations, radiological and operative findings and complications**

| **Patients** | **Number (%)** |
|---|---|
| **Gender** | |
| Male | 9 (60%) |
| Female | 6 (40%) |
| **Median age (range)** | 51 (27–66) years |
| **Fukuhara class** | |
| Class I | 0 |
| Class II | 3 (20%) |
| Class III | 12 (80%) |
| **Mean of duration of symptoms before diagnosis (range)** | 18 (3–45) weeks |
| **Clinical presentations** | |
| Headache | 5 (33.3%) |
| Nausea & vomiting | 1 (6.7%) |
| Gait disturbances | 12 (80%) |
| Cognitive impairment | 8 (53.3%) |
| Urinary incontinence | 6 (40%) |
| **Radiological findings** | |
| Triventricular hydrocephalus due to aqueductal stenosis | 15 (100%) |
| Cortical sulci effacement | 13 (86.6%) |
| Cerebrospinal fluid permeation | 6 (40%) |
| **Operative findings** | |
| Septum pellucidum defect | 1 (6.7%) |
| Second Lilliquist's membrane | 1 (6.7%) |
| Membranous scarring | 2 (13.3%) |
| **Complications** | |
| Transient postoperative fever and/or vomiting | 4 (26.7%) |
| Transient diabetes insipidus | 1 (6.7%) |
| Postoperative meningitis | 1 (6.7%) |

**Table 2: Clinical outcome at the end of the follow-up period**

| Preoperative signs/symptoms | | At the end of the follow-up | | |
|---|---|---|---|---|
| | | Resolved | Improved | Stable |
| Headache | 5 | 1 (20%) | 4 (80%) | 0 |
| Nausea & vomiting | 1 | 1 (100%) | 0 | 0 |
| Gait | 12 | 5 (41.7%) | 7 (58.3%) | 0 |
| Cognitive impairment | 8 | 2 (25%) | 3 (37.5%) | 3 (37.5%) |
| Urinary incontinence | 6 | 0 | 4 (66.7%) | 2 (33.3%) |

## DISCUSSION

According to Fukuhara and Luciano, clinical features of LIAS were classified into 3 different groups of patients in relation to disease duration (Acute, subacute, and chronic LIAS).6 In our study, only two different groups of patients were described; 12 patients presented with chronic clinical history (class III), and 3 cases presented with subacute manifestations (class II). The subacute and chronic LIAS categories were also the most prevalent in Locatelli et al. series, where there were 8 patients in the chronic LIAS group, 4 in the subacute LIAS group, and only one patient in the acute LIAS group.[2]

MRI images of LIAS are characterized by triventricular hydrocephalus with morphologic restrictions or obstruction of the aqueduct of Sylvius. Phase contrast cine MRI is essential for the diagnosis of LIAS in selected patients in whom there are no direct or indirect signs of aqueductal stenosis on the conventional MRI.[14] In our study, conventional preoperative MRI showed evident morphologic stenosis of the aqueduct of Sylvius in all patients, therefore, we did not use phase contrast cine MRI for the diagnosis of LIAS.

All our patients had progressive clinical manifestations which were related to progressive impairment of CSF circulation due to obstruction of the aqueduct of Sylvius and the success rate of ETV among our patients was very high (93.3%), including the patients with either chronic headache or normal pressure hydrocephalus-like manifestations. In our opinion, the improvement of clinical symptoms was achieved by a complete third ventriculostomy which included perforation of the third ventricular floor, Lilliquist's membrane, and basal arachnoid adhesions to achieve not only communication between the third ventricle and the basal cisterns but also a free CSF circulation between the supra-tentorial and infra-tentorial subarachnoid spaces. Similar results were reported by Locatelli et al. in 2014, where ETV was highly successful in the treatment of LIAS in patients with either subacute or chronic clinical manifestations. Similar to our findings, they reported that a clinical history of progressive CSF decompensation is the most essential factor to predict the outcome of ETV in patients with LIAS. They also mentioned that ETV is not only a safe and highly successful procedure in the treatment of patients with LIAS but also it can be considered the treatment of choice in such patients.[2]

As regards the detailed clinical outcome, headache improved in all patients presenting with headache, with complete resolution in one patient. Among 12 patients with gait disturbance, unsteadiness resolved completely in 5 patients and improved partially in 7 patients. Of the 8 patients who presented with cognitive decline, 2 patients resumed their prior social and professional lives, 3 patients showed improvement, and the other 3 patients showed no change. Six patients had urinary incontinence, 4 exhibited improvement, while the remaining 2 patients remained stationary. In Locatelli et al. study, 10 patients presented with gait instability at the time of surgery, in 4 of these patients gait disorder resolved completely, while an improvement was observed in the remaining 6 patients. Cognitive impairment improved in 4 of the 8 patients presenting with cognitive deterioration, one patient returned to their previous professional and social activities, while the other 3 patients showed no change. Urinary incontinence was recorded in seven of their patients, it improved in 4 patients, remained stationary in 2 patients and one patient deteriorated.[2]

There is a considerable debate over the correlation between postoperative ventricular size reduction and a favorable neurological outcome. While some studies supported this hypothesis,[15-18] other studies have failed to demonstrate this correlation.[19-22] Thus, relying on imaging should be avoided because the size of the ventricles is not a reliable indicator of clinical outcomes. In our study, evident ventricular size reduction was obtained in three patients only (20%), however, the majority of our patients improved regardless of the change in the ventricular size, therefore neither the clinical outcome nor the neurological outcome was impacted by this reduction. In a retrospective outcome analysis, Rodis et al. reported that only 5% of patients with chronic LIAS showed a decrease in Evan's ratio after ETV.[4]

Transient postoperative fever and diabetes insipidus were recorded in our series. Only one patient (6.7%) had unsuccessful ETV as he had postoperative meningitis that improved by intravenous and intrathecal antibiotic injection but developed secondary communicating hydrocephalus and required a VP shunt two months later. Locatelli et al. reported no CNS or wound infections in the postoperative period. The ETV was successfully performed in all of their patients and no one required a second ETV procedure or shunt implantation.2 On the other hand, in Fukuhara and Luciano study, 5 of their 31

patients (16.1%) required further CSF diversion. One of these 5 patients experienced intraventricular bleeding during the ETV and required VP shunt placement 1 week following the ETV, while the other 4 patients improved temporarily, requiring VP shunt implantation later.[6]

## CONCLUSION

Endoscopic third ventriculostomy is a safe, effective, and highly successful procedure in the treatment of hydrocephalus due to idiopathic aqueductal stenosis with more than 93.3% success rate. The limitations of this study include the retrospective nature and the small patient population. To enhance our findings, further carefully planned, prospective, multicentric studies on bigger samples should be performed.

### List of Abbreviations

CNS: Central nervous system.
CSF: Cerebrospinal fluid.
CT: Computerized tomography.
ETV: Endoscopic third ventriculostomy.
ICP: Intracranial pressure.
IRB: Institutional review board.
LIAS: Late-onset idiopathic aqueductal stenosis.
MRI: Magnetic resonance imaging.
SPSS: Statistical package for social sciences.
VP: Ventriculoperitoneal.

### Disclosure

The authors report no conflict of interest in the materials or methods used in this study or the findings specified in this manuscript.

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