Childs Nerv Syst (2018) 34:247–255
https://doi.org/10.1007/s00381-017-3626-4

ORIGINAL PAPER

Visual outcome after endoscopic third ventriculostomy for hydrocephalus

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Received: 27 March 2017 / Accepted: 16 October 2017 / Published online: 3 November 2017

Abstract

Purpose Hydrocephalus-related symptoms are mostly improved after successful endoscopic third ventriculostomy (ETV). However, visual symptoms can be different. This study was focused on visual symptoms. We analyzed the magnetic resonance images (MRI) of the orbit and visual outcomes.

Methods From August 2006 to November 2016, 50 patients with hydrocephalus underwent ETV. The male-to-female ratio was 33:17, and the median age was 61 years (range, 5–74 years). There were 18 pediatric and 32 adult patients. Abnormal orbital MRI findings included prominent subarachnoid space around the optic nerves and vertical tortuosity of the optic nerves. We retrospectively analyzed clinical symptoms, causes of hydrocephalus, ETV success score (ETVSS), ETV success rate, ETV complications, orbital MRI findings, and visual impairment score (VIS).

Results The median duration of follow-up was 59 months (range, 3–113 months). The most common symptoms were headache, vomiting, and gait disturbance. Visual symptoms were found in 6 patients (12%). The most common causes of hydrocephalus were posterior fossa tumor in 13 patients, pineal tumor in 12, aqueductal stenosis in 8, thalamic malignant glioma in 7, and tectal glioma in 4. ETVSS was 70 in 3 patients, 80 in 34 patients, and 90 in 13 patients. ETV success rate was 80%. ETVSS 70 showed the trend in short-term survival compared to ETVSS 90 and 80. ETV complications included epidural hematoma requiring operation in one patient, transient hemiparesis in two patients, and infection in two patients. Preoperative abnormal orbital MRI findings were found in 18 patients and postoperative findings in 7 patients. Four of six patients with visual symptoms had abnormal MR findings. Three patients did not show VIS improvement, including two with severe visual symptoms.

Conclusions Patients with severe visual impairment were found to have bad outcomes. The visual symptoms related with increased intracranial pressure should be carefully monitored and controlled to improve outcomes.

Keywords Hydrocephalus · Endoscopic third ventriculostomy · Visual outcome · Orbital MRI findings

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Introduction

Endoscopic third ventriculostomy (ETV) is a treatment modality for hydrocephalus. It is the treatment of choice for non-communicating hydrocephalus. A wide range of success rates of endoscopic management for hydrocephalus have been reported: from 30 to 100% [1–4]. Hydrocephalus-related symptoms are mostly improved after successful ETV. However, visual symptoms can be different. After increased intracranial pressure, the direct transmission of cerebrospinal fluid to the orbital cavity causes papilledema and visual disturbance [5]. Hydrocephalus can cause multiple ophthalmic abnormalities and sometimes life-threatening conditions with severe visual

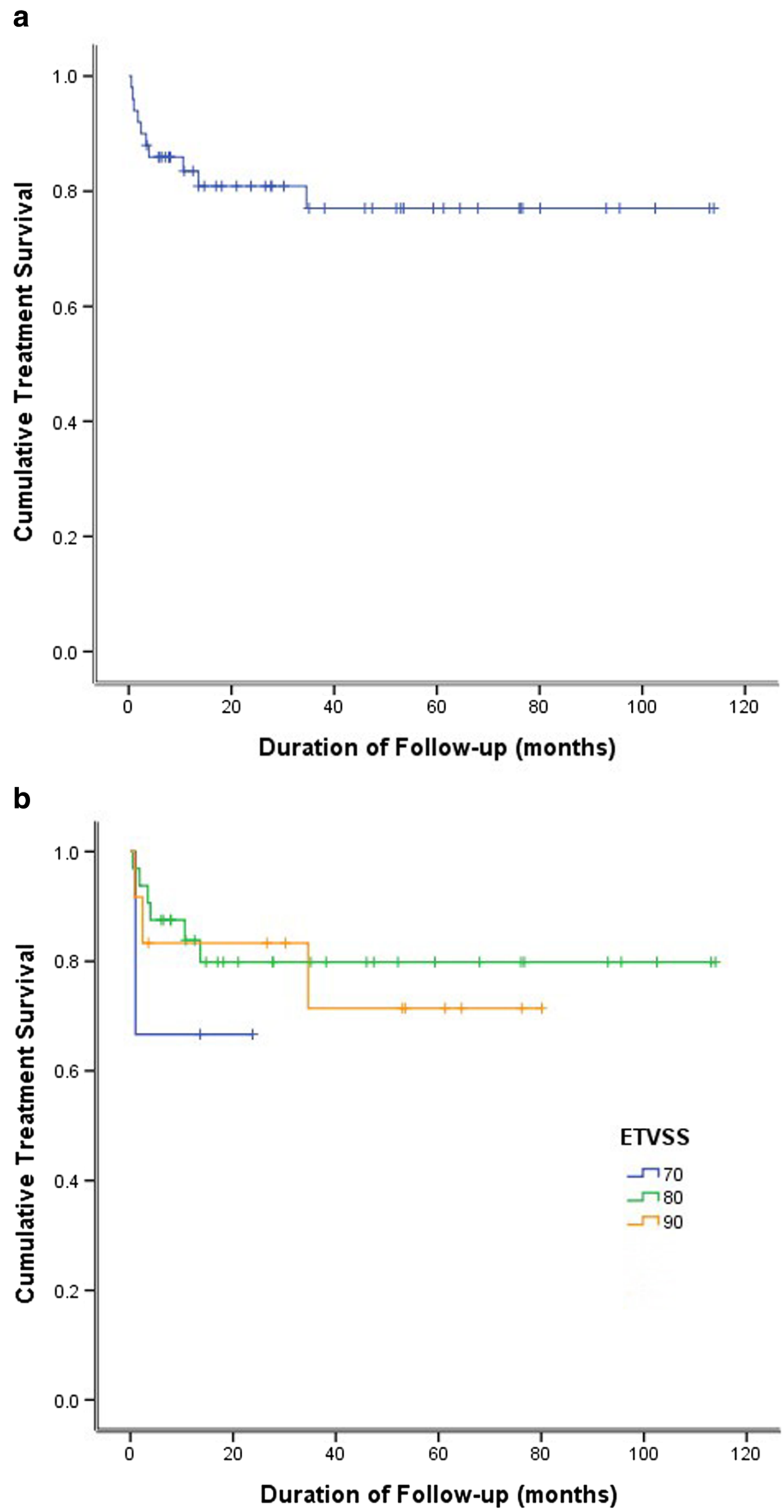

Fig. 1 The treatment survival of ETV on Kaplan-Meier curve. **a** The mean treatment survival of ETV was 90.0 (± 6.8) months on the time course of failure. **b** Depending on ETVSS, the mean treatment survival was 61.6 (± 9.2) months for ETVSS 90, 92.2 (± 8.0) months for 80, and 16.2 (± 6.2) months for 70 ($p = 0.717$). ETVSS 70 showed the trend in short-term survival among them

symptoms [6–9]. Some patients had permanent visual loss even after successful control of intracranial pressure.

ETV is a surgical procedure in which an opening is created in the floor of the third ventricle using an endoscope placed

Table 1 The relation between visual symptoms and orbital MRI findings

Preoperative and postoperative radiologic and clinical findings	Abnormal orbital findings (width of ONS, mean ± SD, mm)	No abnormal orbital findings (width of ONS, mean ± SD, mm)	*p* value
Before ETV (*N* = 50)			
Preop. visual symptoms	*N* = 4 (7.7 ± 0.5)	*N* = 2 (5.9 ± 0.3)	0.07
Preop. no visual symptoms	*N* = 14 (7.55 ± 0.4)	*N* = 30 (5.7 ± 0.5)	
Successful ETV (*N* = 40)			
Postop. visual symptoms	*N* = 1 (7.3)	*N* = 1 (5.3)	0.28
Postop. no visual symptoms	*N* = 2 (7.1 ± 0.1)	*N* = 36 (5.3 ± 0.7)	
Failed ETV (*N* = 10)			
Postop. visual symptoms	*N* = 1 (7.3)	*N* = 0	0.60
Postop. no visual symptoms	*N* = 3 (7.3 ± 0.2)	*N* = 6 (5.3 ± 0.8)	

ETV endoscopic third ventriculostomy, *Orbital findings* orbital MRI findings, *ONS* optic nerve sheath, *p value* statistical significance between visual symptoms and the width of ONS

within the ventricular system through a burr hole. In contrast to ventriculoperitoneal shunt, ETV requires a longer time to control increased intracranial pressure with a small intracranial opening. In patients with acute and severe visual symptoms, ETV cannot effectively restore the visual symptoms within a short time. This study focused on visual symptoms caused by hydrocephalus. We analyzed the magnetic resonance images (MRI) of the orbit and patients' visual symptoms before and after ETV.

Methods

Patients' clinical characteristics

From August 2006 to December 2016, 50 patients with hydrocephalus underwent ETV. The male-to-female ratio was 33:17, and the median age was 61 years (range, 5–74 years). The median duration of follow-up was 59 months (range, 3–113). There were 18 pediatric and 32 adult patients. We retrospectively analyzed clinical symptoms, causes of hydrocephalus, ETV success score (ETVSS), ETV success rate, ETV complications, orbital MRI findings, and visual impairment score (VIS).

The chief clinical symptoms were headache in 18 patients, vomiting and headache in 9, gait disturbance in 11, drowsy mental state in 4, dizziness in 5, memory impairment in 2, and seizure in 1. Six patients out of 50 (12%) had associated visual symptoms. The etiology of hydrocephalus was posterior fossa tumor in 13 patients, pineal tumor in 12, aqueductal stenosis in 8, thalamic malignant glioma in 7, tectal glioma in 4, thalamic neuroglial cyst in 1, third ventricle neurocysticercosis in 1, arteriovenous malformation with hemorrhage in 1, subependymal giant cell astrocytoma on lateral ventricle in 1, petroclival meningioma in 1, and trigeminal schwannoma in 1. There was no case with lesions along the optic pathway. The type of hydrocephalus was obstructing in 49 patients, and communicating in 1 patient. There was no patient with previous shunt history.

Table 2 Six cases with visual symptoms

Cases	Before ETV				After ETV			Outcome	
	Duration of VS	Width of ONS (mm)	VIS (VA)	VIS (VFD)	Width of ONS (mm)	VIS (VA)	VIS (VFD)	Result of ETV	Clinical visual improvement
1	8 months	5.6	10	26	5.3	10	26	Success	Not improved
2	Several years	7.3	20	nc	7.3	30	nc	Fail - > success after re-ETV	Not improved after success of re-ETV
3	Several years	8.2	92	25	7.3	92	25	Success	Not improved
4	2 months	7.1	37	nc	7.2	27	nc	Success	Improved
5	4 months	8.0	4	25	4.8	4	2	Success	Improved
6	2 months	6.2	47	nc	4.5	4	nc	Success	improved

ETV endoscopic third ventriculostomy, *VS* visual symptoms, *ONS* optic nerve sheath, *VIS* visual impairment score, *VA* visual acuity, *VFD* visual field defect, *nc* not checked

Analysis of ETVSS, visual symptoms, and orbital MRI findings

ETVSS is based on age, etiology of the hydrocephalus, and presence of a previous shunt [10]. The ETVSS is calculated as age score (< 1 month, 0; 1 month to < 6 months, 10; 6 months to < 1 year, 30; 1 year to < 10 years, 40; ≥ 10 years, 50) + etiology score (postinfectious, 0; myelomeningocele, intraventricular hemorrhage, non-tectal brain tumor, 20; aqueductal stenosis, tectal tumor, other, 30) + previous shunt score (previous shunt, 0; no previous shunt, 10). ETVSS ranges from 0 (extremely poor chance of ETV success) to 90 (extremely high chance of success).

The success criteria of ETV was defined as a normalized or improved clinical state and at least one of the following radiologic criteria: (a) reduction in ventricular size or stable ventricles with disappearance of periventricular edema and an increase in subarachnoid space over cerebral convexities, or (b) flow artifact in sagittal T2 fast spin echo magnetic resonance images.

Abnormal orbital MRI findings included prominent subarachnoid space around the optic nerves and vertical tortuosity of the optic nerves. The width of the optic nerve sheath (ONS) directly behind the globe was above 7.0 mm on abnormal MR imaging [11]. Ophthalmological examination included visual acuity and field charting. The findings were analyzed according to the guidelines of the German Ophthalmological Society [12]. The scores for visual acuity and visual field defects in each patient were added to provide the visual impairment score (VIS), which enabled an exact comparison between different examinations in each patient. The VIS ranges from 0 to 100.

Three months was the minimum postoperative follow-up required in all patients for evaluation of ETV success and visual outcome.

Statistical analysis

We calculated the treatment survival of ETV by the Kaplan-Meier method and analyzed the influence of ETVSS on the treatment survival by log rank rest. Treatment survival was calculated from the date of surgery to the date of treatment failure or last follow-up. A *t* test was performed to analyze the statistical difference between visual symptoms and the width of ONS. All statistical analyses were performed at a significance level of $p < 0.05$ using the statistical package SPSS 21.0 (SPSS Inc., Chicago, IL, USA).

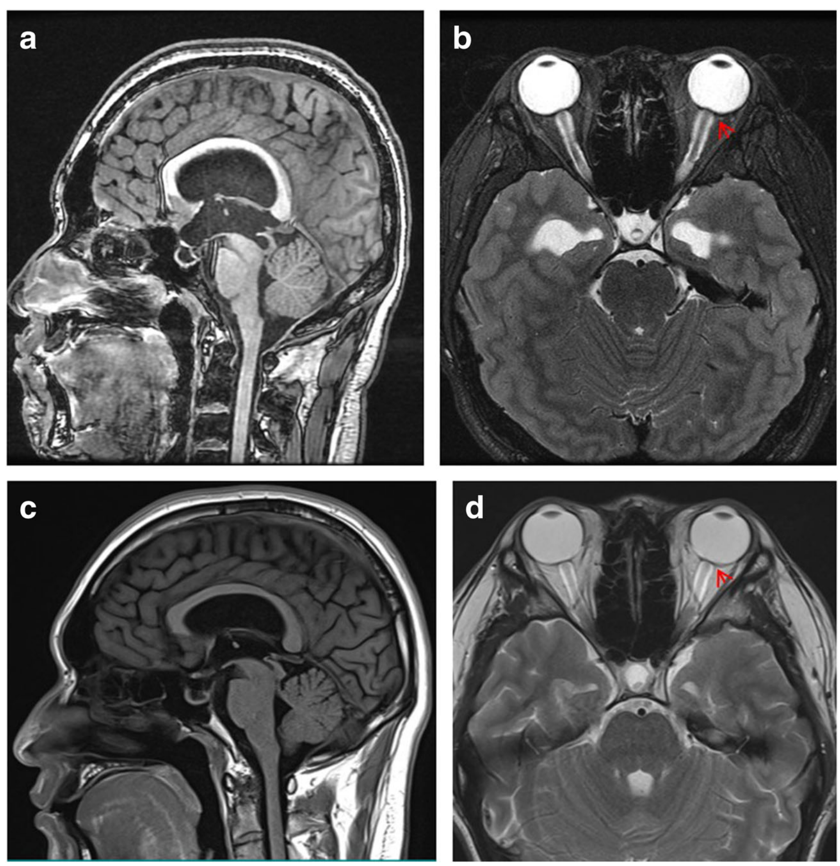

Fig. 2 Radiologic findings of case 3 with no change of severe visual symptoms after ETV. **a** Obstructing hydrocephalus due to aqueductal stenosis on T1-weighted sagittal MRI. **b** Prominent subarachnoid space (red arrow) around the optic nerve on T2-weighted axial MRI. The width of the optic nerve sheath directly behind the globe was 8.2 mm. **c** Six months after ETV, decreased ventricular volume can be visualized on T1-weighted sagittal MRI. **d** Six months after ETV, the remaining subarachnoid space (red arrow) is visualized on T2-weighted MRI. The width of the optic nerve sheath was 7.3 mm

Results

Outcome of endoscopic third ventriculostomy

ETV success rate was 80%. ETVSS was 70, 80, and 90 in 3, 34, and 13 patients, respectively. There were 10 failure cases, 3 thalamic malignant tumors, 2 aqueductal stenosis, 1 petroclival meningioma, 1 trigeminal schwannoma, 1 pineal tumor, 1 neurocysticercosis, and 1 arachnoid cyst on the quadrigeminal cistern. A second ETV was performed in two cases of aqueductal stenosis and one case of arachnoid cyst in the quadrigeminal cistern, which were successful. A ventriculoperitoneal shunting was performed in four cases, two thalamic malignant tumors, one trigeminal schwannoma, and one pineal tumor. Three patients gave up further treatment.

The mean treatment survival was 90.0 (± 6.8) months. Figure 1a showed the time course of failure on Kaplan-Meier curve. Depending on ETVSS, the mean treatment survival was 61.6 (± 9.2) months for ETVSS 90, 92.2 (± 8.0) months for 80, and 16.2 (± 6.2) months for 70. ETVSS 70 showed the trend in short-term survival among them. There was no statistical significance (Fig. 1b, $p = 0.717$). ETV complication rate was 8%. The complications were epidural hematoma requiring operation in one, transient hemiparesis in two, and meningitis in one patient.

Visual symptoms and orbital MRI findings before and after endoscopic third ventriculostomy

The relation between visual symptoms and orbital MRI findings were summarized in Table 1. Preoperative abnormal orbital MRI findings were seen in 18 out of 50 patients (36%). Out of the 18 patients, 4 patients (22%) complained of visual symptoms. Postoperatively, abnormal orbital MRI findings were seen in 7 out of 50 patients (14%), and 2 out of the 7 (29%) patients had visual symptoms. The width of ONS was summarized in Table 1. There was no statistical significance between

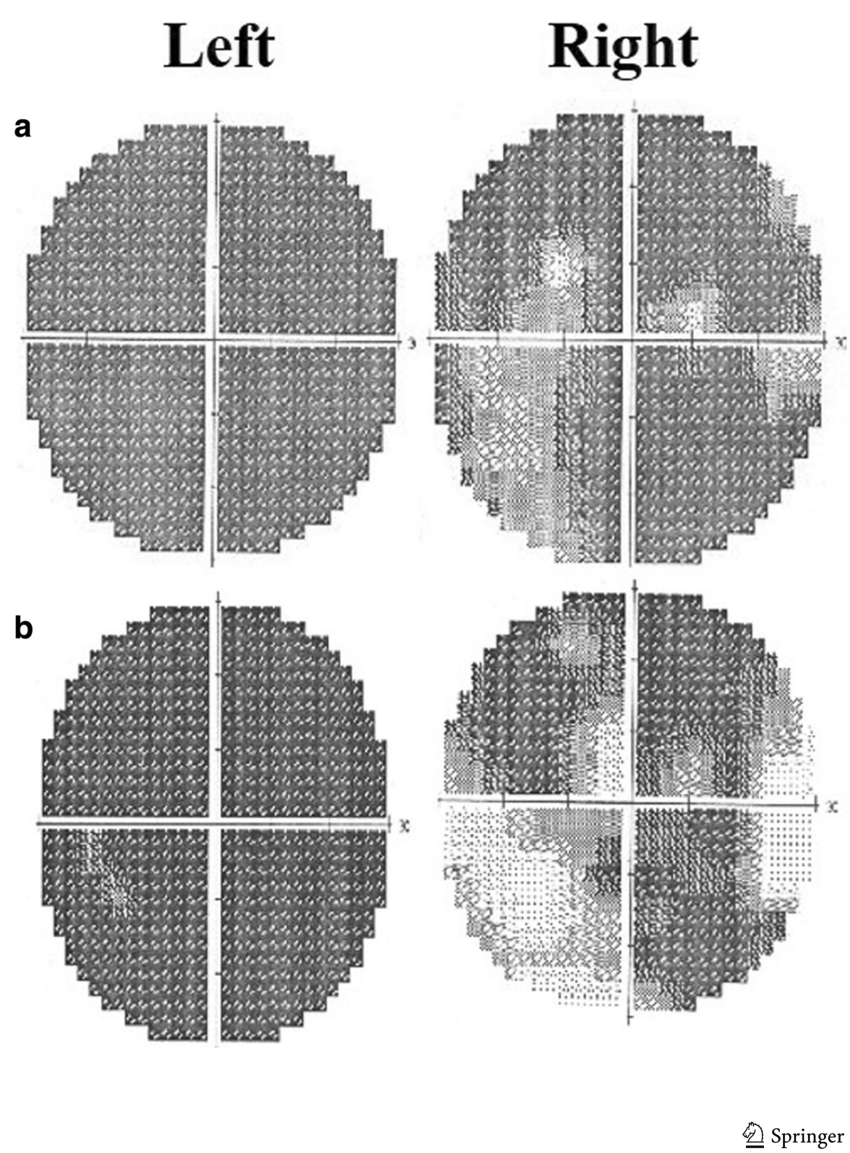

Fig. 3 Visual field of case 3 with no change of severe visual symptoms after ETV. **a** Preoperatively, there was an impaired visual field except in the right medial inferior area. **b** Six months after surgery, there was mostly an impaired visual field

visual symptoms and the width of ONS before and after ETV.

Out of all 50 patients, 6 patients (12%) complained of visual symptoms preoperatively. Six cases with visual symptoms were summarized in Table 2. ETV was successful in six patients including one having re-ETV. Preoperatively, four patients had abnormal orbital MRI findings, and three postoperatively retained these abnormal findings. VIS was evaluated in these patients. Postoperatively, three patients did not show VIS improvement including two patients who complained of preoperative severe visual symptoms and were found to have postoperative abnormal MRI findings.

Illustration of case 3: no change of severe visual symptoms after ETV

An 18-year-old male patient complained of headache and visual disturbance. Visual acuity had been worsening for 3 months. Preoperative MRI showed an obstructing hydrocephalus associated with aqueductal stenosis and a prominent subarachnoid space around the optic nerves (Fig. 2a, b). ETV was performed. Postoperatively, headache was improved without a change in visual symptoms. After 6 months, brain MRI showed decreased ventricular volume with the remaining subarachnoid space around the optic nerves (Fig. 2c, d). Preoperatively, visual acuity was 0.08 on the right eye and 0.02 on the left. His visual field was mostly impaired (Fig. 3a). Preoperative VIS was 100. The postoperative visual acuity, visual field, and VIS were the same as the preoperative values (Fig. 3b).

Illustration of case 5: visual improvement after ETV

A 51-year-old woman complained of headache and visual disturbance. Headache and visual symptoms had been persistent for 4 months. Preoperative MRI showed an obstructing hydrocephalus with thalamic glial cyst and the subarachnoid space around the optic nerves (Fig. 4a, b). ETV was performed. Postoperatively, headache and visual disturbance were improved. Nine months later, brain MRI showed decreased ventricular volume and subarachnoid space around the optic nerves (Fig. 4c, d). Preoperative visual acuity was 0.9 on the right eye and 0.8 on the left eye; the visual field was mostly impaired (Fig. 5a). Preoperative VIS was 29.

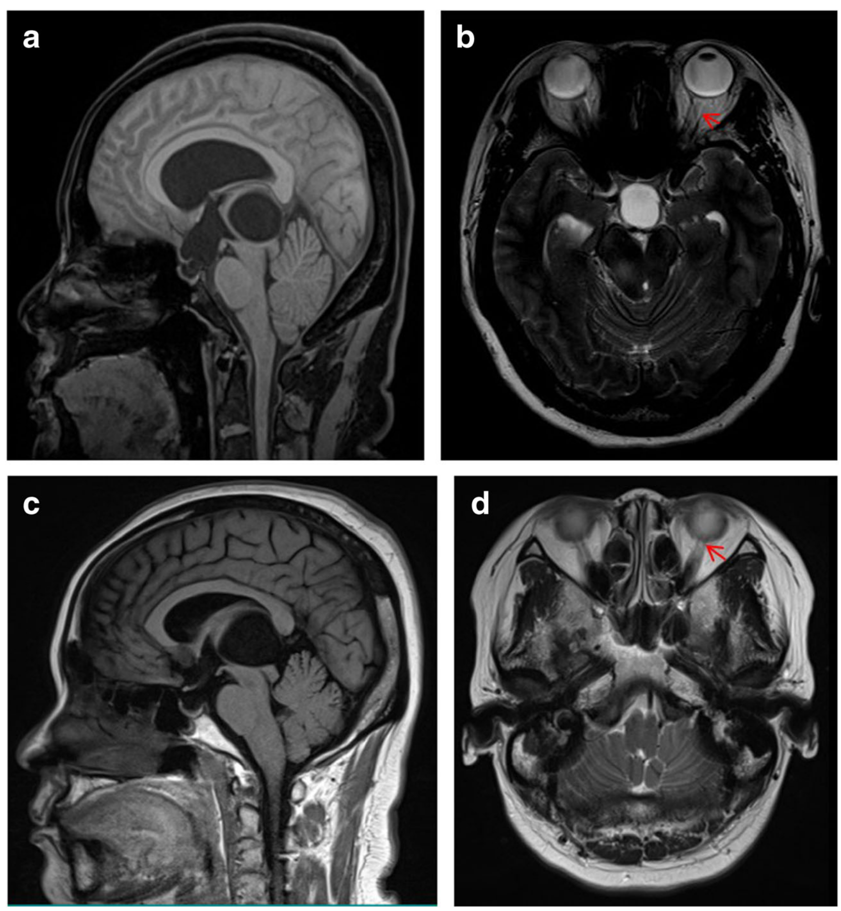

Fig. 4 Radiologic findings of case 5 with visual improvement after ETV. **a** Obstructing hydrocephalus with a thalamic glial cyst on T1-weighted sagittal MRI. **b** Prominent subarachnoid space (red arrow) around the optic nerve on T2-weighted axial MRI. The width of the optic nerve sheath directly behind the globe was 8.0 mm. **c** Six months after ETV, decreased ventricular volume can be visualized on T1-weighted sagittal image. **d** Six months after ETV, no subarachnoid space (red arrow) is visualized on T2-weighted MR image. The width of the optic nerve sheath was 4.8 mm

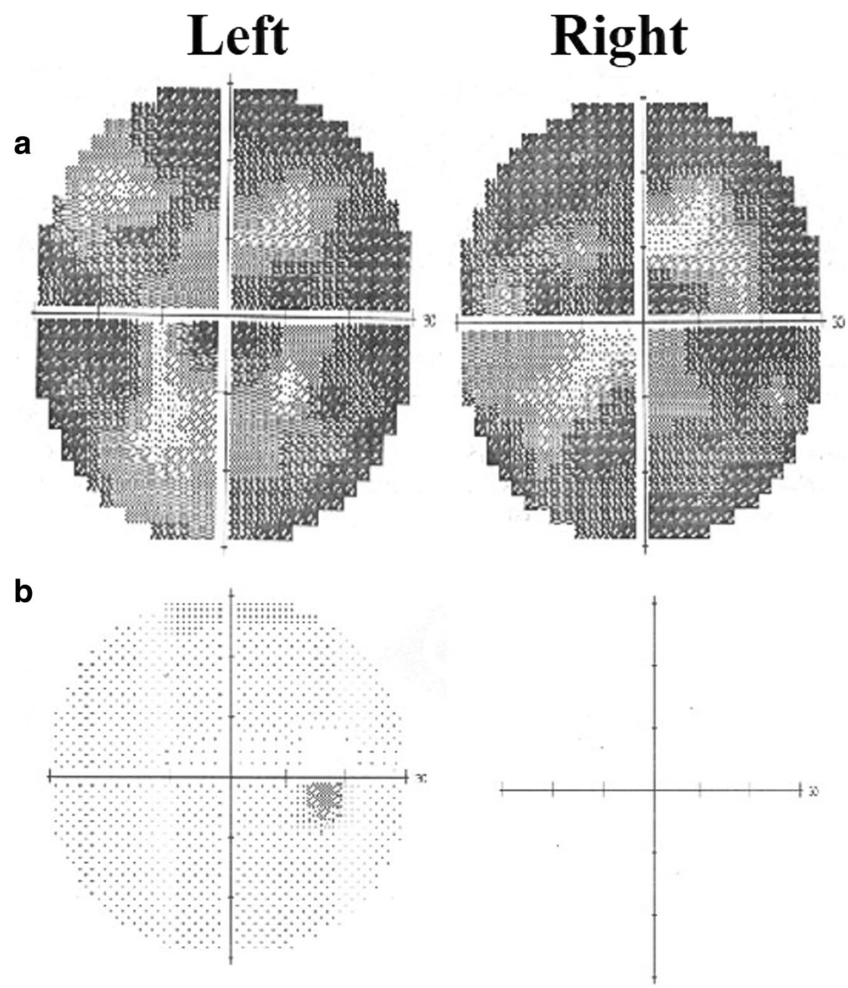

Fig. 5 Visual field of case 5 with visual improvement after ETV. **a** Preoperatively, there was an impaired visual field. **b** Six months after surgery, there was an improved visual field

Postoperatively, visual acuity was 0.9 on both eyes, and the visual field was improved (Fig. 5b). Postoperative VIS was 6.

Discussion

An ETV is the internal diversion of cerebrospinal fluid through the floor of the third ventricle. It is an important treatment tool for obstructing hydrocephalus. Even if the early failure rate is high, a successful ETV can provide a long-term treatment effect without complications of cerebrospinal fluid shunting. Previous studies reported that age, cause of hydrocephalus, and history of previous cerebrospinal fluid shunting are independent factors that predict success of ETV [2, 4, 10]. As the cause of hydrocephalus, non-tectal brain tumor showed the higher failure rates compared to tumor of the midbrain tectum [2]. In this study, six patients had non-tectal brain tumor out of 10 ETV failure cases. Four patients with thalamic malignant glioma and pineal tumor showed delayed ETV failure with tumor recurrence, and two patients with trigeminal schwannoma and petroclival meningioma showed early failure.

Increased intracranial pressure from hydrocephalus causes headache, nausea, gait disturbance, mental status changes, memory impairment, and visual disturbance. Ophthalmological abnormalities from hydrocephalus such as strabismus, amblyopia, decreased visual acuity, refractive errors, abnormal ocular motility, and visual field constriction have been reported in 18 to 60% of patients [13]. Severe visual symptoms such as acute visual loss associated with malfunction of cerebrospinal fluid shunting have been reported [9, 14]. In hydrocephalic patients with acute or aggravated visual symptoms, it is necessary to suspect increased intracranial pressure. The fragility of the visual system such as abnormal optic disc and retinal vessels may cause rapid deterioration of visual symptoms in patients with hydrocephalus, especially in children [7]. Children with shunt-treated hydrocephalus showed the concentric visual field constriction in 57%, and they could have a higher risk of having ophthalmological abnormalities [11]. Elevated ONS pressure may not be relieved after surgical procedures to decrease intracranial pressure for hydrocephalus [15, 16]. This could be explained as relatively increased fluid flow into the orbit even after decreased intracranial pressure.

Hydrocephalus can cause papilledema and visual disturbance due to axoplasmic stasis and distention of the optic nerve because of the direct transmission of cerebrospinal fluid pressure to the optic cavity [5]. Reported MRI findings include the dilation of the subarachnoid space around the optic nerve, flattening of the posterior sclera, and vertical tortuosity of the orbital optic nerve [17, 18]. Edema in the optic chiasm can be frequently observed in obstructing hydrocephalus, which is reversible [19]. These orbital MRI findings may indicate increased intracranial pressure [20]. However, these findings may not be associated with clinical visual symptoms [19, 20]. In this study, abnormal orbital MRI findings were observed in 18 out of 50 patients (36%). And these were reversible in 11 patients after ETV. The ONS diameter is a strong indicator of increased intracranial pressure [11]. However, the correlation between the width of ONS and visual disturbance was not evident in this study.

After resolution of hydrocephalus, most related symptoms are improved. However, permanent visual loss has been reported even though increased intracranial pressure was controlled [8, 9]. After ETV, a small opening is made in the floor of the third ventricle, allowing the flow of cerebrospinal fluid into the normal space of the interpeduncular cistern. This intracranial cerebrospinal fluid diversion normalizes intracranial pressure without using mechanical shunting. Although ETV is an ideal physiologic procedure, patients may not maintain adequate cerebrospinal fluid to normalize intracranial pressure immediately after the procedure. In contrast to cranial shunting, cerebrospinal fluid reabsorption pathways require some time to accommodate the increased amount of cerebrospinal fluid. In this study, three out of six patients with visual symptoms did not have visual improvement postoperatively, including two patients who complained of severe visual symptoms preoperatively. Early detection and treatment of hydrocephalus may allow the recovery of visual function. Neurosurgeons have to control intracranial pressure after ETV in cases of acute and severe visual symptoms. Postoperatively, external ventricular drainage could be helpful in controlling increased intracranial pressure to improve acute symptoms. The duration of external ventricular drainage could be flexible depending on related symptoms, and shunting may be considered as an alternative treatment for long-standing symptoms.

Conclusions

Patients with severe visual impairment showed poor outcomes. The visual symptoms related with increased intracranial pressure should be carefully monitored and controlled to improve outcomes.

Acknowledgements This study was supported by the Basic Science Research Program through the National Research Foundation of Korea (NRF), funded by the Ministry of Science, ICT, & Future Planning (2017R1A1A1A05001020).

Compliance with ethical standards

Conflict of interest The authors declare no conflicts of interest.

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