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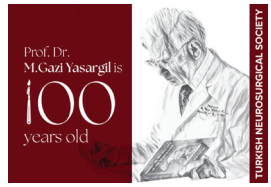

Proactive External Lumbar Drainage Use in Pediatric Idiopathic Intracranial Hypertension and Proposal of a New Treatment Algorithm

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ABSTRACT

AIM: To determine if the concurrent use of external lumbar drainage (ELD) and oral medication will hasten the decrease in intracranial pressure (ICP) and resolution of papilledema in pediatric idiopathic intracranial hypertension (IIH).

MATERIAL and METHODS: In this retrospective study, we evaluated the outcome of pediatric patients with IIH who underwent ELD as an adjunct treatment to standard oral medications. All patients underwent ophthalmological examination, optic coherence tomography, retinal nerve fiber layer thickness assessment, and ICP measurements before and after ELD. The outcome was evaluated via serial ophthalmological examinations, optical coherence tomography to measure retinal nerve fiber layer thickness, and lumbar puncture to measure ICP.

RESULTS: Eleven pediatric patients (7 females, 4 males) were enrolled in the study. The mean age of the patients was 10.9 ± 4.4 years (range, 5.6–17.7 years). The mean cerebrospinal fluid opening pressure was 447 ± 112.5 mm H_2O before ELD. The mean post-ELD ICP was 263.1 ± 92.4 mm H_2O. The retinal nerve fiber layer thickness at the time of diagnosis was 200.9 ± 113.7 µm and 212.6 ± 123.3 µm in the right and left eyes, respectively. After ELD, the thickness was 149.4 ± 45 µm and 151.4 ± 51.3 µm in the right and left eyes, respectively. The mean duration of ELD was 8.7 ± 1.4 days (range, 7–10 days). The post-ELD cerebrospinal fluid opening pressure and retinal nerve fiber layer thickness were significantly lower than pre-ELD values. Four patients required lumboperitoneal shunt surgery during follow-up.

CONCLUSION: Proactive ELD is an effective method to achieve a rapid decrease in ICP and retinal nerve fiber layer thickness without major complications.

KEYWORDS: Papilledema, Intracranial hypertension, Lumbar drainage, Optical coherence tomography, Pediatric pseudotumor cerebri

ABBREVIATIONS: IIH: Idiopathic intracranial hypertension, **ICP:** Intracranial pressure, **CSF:** Cerebrospinal fluid, **OCT:** Optic coherence tomography, **LPS:** Lumboperitoneal shunt, **ONSF:** Optic nerve sheet fenestration, **ELD:** External lumbar drainage, **LP:** Lumbar puncture

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INTRODUCTION

Idiopathic intracranial hypertension (IIH) is defined as an increase in intracranial pressure (ICP) in the absence of an intracranial mass lesion that is associated with normal-sized or slit ventricles and normal cerebrospinal fluid (CSF) biochemistry (8). IIH is a rare disease with an estimated incidence of 6–9 per 1,000,000 people in the pediatric population (7,9), and its incidence increases with obesity (10). The common presenting symptoms of IIH are headache, nausea, vomiting, blurred vision, double vision secondary to 6th nerve palsy, and transient visual deficits (1,17).

The most serious complication of IIH is visual loss (8-10,16), with a frequency of approximately 50% (6,27,28). Therefore, a reliable ophthalmological examination is mandatory during the diagnosis and follow-up of IIH. Fundus examination may sometimes be challenging, especially in subtle papilledema, which may be present in early-onset IIH (22). Optical coherence tomography (OCT) is a reliable and objective tool for diagnosing and monitoring optic nerve disorders, including papilledema (22). Furthermore, it is useful for measuring optic disk swelling, even in young children (7).

The pathology underlying IIH is hypothesized to be venous sinus hypertension, which decreases CSF absorption and leads to increased ICP (10). Although oral medications that decrease CSF production are widely used, surgical treatment options, such as lumbo-peritoneal shunt (LPS), optic nerve sheath fenestration (ONSF), and venous sinus stenting, are available for patients with refractory IIH (3,4). In recent studies, external lumbar drainage (ELD) has been used in pediatric patients with IIH in whom oral medications have failed (7,10,14).

In of our study, we aimed to investigate whether the combined use of ELD and oral medications at the time of diagnosis could provide a faster reduction in ICP and reduce the risk of optic nerve damage in pediatric patients with IIH. This proactive approach was designed to avoid the need for early surgical shunting, and thereby potentially reducing long-term optic nerve damage. Previous studies have demonstrated that delaying surgical intervention may increase the risk of irreversible visual damage. This justifies our approach of rapid intervention with ELD and medication. We hypothesized that the combined use of ELD and oral medication could be a more effective and less invasive first-line treatment strategy than early direct shunting.

MATERIAL and METHODS

In this retrospective study, we analyzed the medical records of pediatric patients diagnosed with IIH in our Pediatric Neurosurgery Clinic between February 2020 and February 2022. This study was approved by the institutional ethics committee, and informed consent was obtained from the legal guardians of all the patients (No: 2025-03/127). All the methods were performed in accordance with the relevant guidelines and regulations. Patients who underwent ELD during their IIH treatment were included in the study. All patients with signs and symptoms of IIH, including papilledema during a routine ophthalmological examination, visual disturbances, headache, and vomiting, were examined by the pediatric IIH study group. The group consisted of a pediatric neurologist, pediatric ophthalmologist, pediatric neurosurgeon, pediatric neuroradiologist, and nurse practitioner. All patients underwent thorough ophthalmological and neurological examinations, cranial 3T magnetic resonance imaging (MRI) to exclude hydrocephalus and any intracranial mass lesion, magnetic resonance venography with contrast to evaluate the venous sinus anatomy, OCT (Zeiss), and lumbar puncture (LP). In accordance with Friedman's criteria, an CSF opening pressure of >280 mm H_2O in obese patients or >250 mm H_2O in nonobese patients was accepted as high. All patients with an CSF opening pressure of >300 mm H_2O underwent ELD at the same time as the LP. According to our pediatric IIH treatment algorithm, all legal guardians had been informed of the concurrent ELD insertion if the CSF opening pressure was >300 mm H_2O, and informed consent was obtained (Figure 1).

All LP and ELD procedures were performed by the same neurosurgeon in an operating room under sedation (without endotracheal intubation) with anesthetic agents that did not increase the ICP (e.g., propofol and remifentanil). The obtained CSF was examined for cell count, protein and glucose levels, and bacterial growth in every patient. After the ELD was inserted, 10–15 cc/h of CSF was drained for at least 5 days. The ICP was concomitantly monitored (IntelliVue MX500; Philips) via the ELD kit, and the exact drainage amount was tailored according to the ICP. Furthermore, the retinal nerve fiber layer (RNFL) thickness was measured in each patient via OCT. All patients were started on oral medications on the first day of the ELD. Non-obese patients were administered acetazolamide, and obese patients were administered topiramate (TPM) (Table I).

The OCT and ophthalmological examination were repeated on the fifth day. If the papilledema had not decreased, ELD was continued. Thereafter, ophthalmological examination and OCT were repeated on days 7 and 10 of ELD. The ELD kit was removed at the end of the 10th day to avoid the increased risk of contamination (20). Before ELD removal, the last ICP pressure (obtained 24 h after clamping the ELD) was noted for each patient. If this value was >280 mm H_2O and/or there was no improvement in papilledema, an LPS system was implanted.

All patients who benefited from the oral medications and ELD were followed up every 6 weeks. During the follow-up, the CSF pressure was measured via LP and neurological and ophthalmological examinations were performed. OCT was repeated during two visits. Routine follow-up was scheduled for 3 months, 6 months, and 1 year accordingly.

The results were analyzed using IBM SPSS (version 20.0, USA). Descriptive statistics are reported as mean ± standard deviation. The mean OCT-RNFL thickness was analyzed using paired t-test. All statistical tests were two-tailed, and a p-value of <0.05 was considered statistically significant.

Table I: Summary of Demographic and Clinical Data of Patient Cohort

Patient	Age (yrs.)	Sex	BMI	MRI	CSF Pressure (mmH_2O)	Complaint	Treatment	CSF Pressure (Post ELD) (mmH_2O)	RNFL (at presentation) (R-L)	RNFL (Post ELD) (R-L)	LP Shunt	Follow-up period (mo)	Outcome
1	5.8	F	N	PES, OSD	350	No	ACT+ELD (10 days)	340	310/365	249/278	Yes (2 weeks after ELD)	25.1	Uneventful
2	6.8	F	OW	N	320	No	ACT+ELD (7 days)	190	112/138	109/129	No	5.2	Uneventful
3	10	F	OB	PES, OSD	430	No	TPMX + ELD (7 days)	180	113/113	110/108	No	5.8	Uneventful
4	17.7	F	UW	N	380	No	ACT+ELD (7 days)	140	130/131	124/114	No	8.2	Uneventful
5	10.9	F	OB	PES, OSD	550	H/A, vomiting	ACT+ELD (10 days)	195	231/294	140/165	No	16.5	Uneventful
6	17.5	F	OB	PES, OSD	510	H/A, eye pain	TPMX + ELD (10 days)	210	140/143	146/135	Yes (4 weeks after ELD)	4	Uneventful
7	14.7	M	OW	PES, OSD+ON kinking	450	Precocious Puberty	TPMX + ELD (7 days)	260	234/253	157/153	No	16.9	Uneventful
8	12	M	OB	PES, OSD	650	H/A, 6^{th} palsy	ACT+ELD (10 days)	420	485/484	223/215	Yes (just after ELD)	20	Uneventful
9	5.6	M	OW	PES, OSD	450	H/A, nausea, vomiting	ACT+ELD (8 days)	400	195/175	139/143	Yes (just after ELD)	16.3	Uneventful
10	6.4	M	N	PES, OSD	360	6^{th} palsy	ACT+ELD (10 days)	300	121/115	117/112	No	2	Uneventful
11	12.8	F	OW	PES, OSD	470	H/A	ACT+ELD (10 days)	260	139/128	130/114	No	2	Uneventful
Mean	10.9	--	--	--	447	--	--	263.1 ($p<0.01$)	200.9/212.6	149.4/151.4 ($p<0.05$)			

ACT: *Acetazolamide;* ***CSF:*** *Cerebrospinal fluid;* ***ELD:*** *external lumbar drainage;* ***H/A:*** *headache;* ***L:*** *left;* ***LP:*** *lumboperitoneal;* ***mo:*** *months;* ***N:*** *normal;* ***OB:*** *Obese;* ***OSD:*** *Optic nerve sheath dilatation,* ***OW:*** *Over weight;* ***UW:*** *Underweight;* ***PES:*** *Partial empty sella;* ***RNFL:*** *Retinal nerve fiber layer thickness;* ***TPMX:*** *Topiramate;* ***yrs:*** *Years*

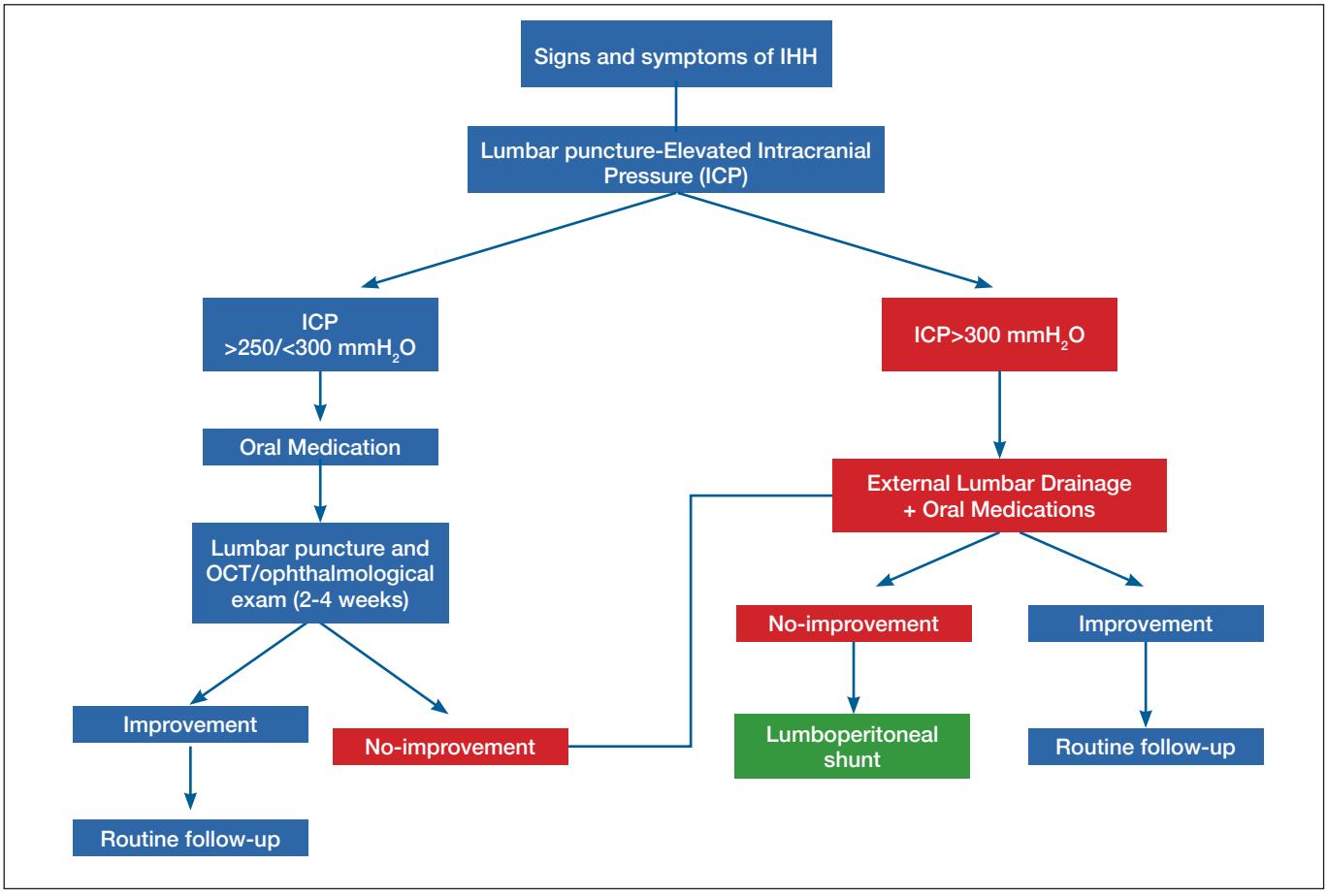

Figure 1: Our pediatric idiopathic intracranial hypertension patient management algorithm.

RESULTS

The study included 11 patients (7 females and 4 males). The mean age at the time of diagnosis was 10.9 ± 4.4 years (range, 5.6–17.7 years). The mean follow-up period was 11.1 ± 8.1 months (range, 2–25.1 months). Five children presented with headaches, and four of them had additional presentations such as lateral gaze palsy and double vision, nausea, vomiting, or pain in the eye. Among the remaining 6 children who did not have headache, one had isolated right lateral gaze palsy and one had precocious puberty. None of the patients had concomitant venous sinus thrombosis. However, one child had a history of venous sinus thrombosis 7 months ago that had resolved. This child and four other children had no complaints, and only bilateral papilledema had been detected in them during the yearly ophthalmological examinations. Bilateral papilledema had been detected in all the study participants during an ophthalmological examination at admission. Body mass index were interpreted according to the age–sex-matched percentiles of Turkish children (5). One patient was underweight (female), two patients had normal weight (1 female and 1 male), four patients were overweight (2 female and 2 male), and four patients were obese (3 female and 1 male).

In eight patients, the MRIs demonstrated increased CSF content within the optic nerve sheath and a partially empty sella. In the remaining three patients, the MRIs were completely normal.

Eight patients were administered acetazolamide, and three patients were administered TPM. These medications were concurrently started with ELD insertion and followed-up by the same pediatric neurologist. The medications were continued after ELD withdrawal in accordance with our pediatric IIH treatment algorithm. The demographic and clinical data of our cohort are summarized in Table I.

The mean CSF opening pressure at the time of diagnosis was 447 ± 97 mm H_2O. The mean CSF opening pressure was 477.5 ± 112.5 mm H_2O in males and 430 ± 85 mm H_2O in females. The mean post-ELD (just before ELD removal) CSF pressure was 263.1 ± 92.4 H_2O. The mean post-ELD CSF pressure was 345 ± 77.2 mm H_2O in males and 216.4 ± 65.2 mm H_2O in females. The CSF pressures had significantly decreased after ELD insertion ($p<0.01$). There was no significant difference in the pre- and post-ELD mean CSF opening pressures between the two sexes. A few patients underwent a post-ELD MRI with contrast, which showed an increase in the diameter of the transverse sinuses (patients 5 and 10).

The mean OCT-RNF thickness at the time of diagnosis was 200.9 ± 113.7 µm in the right eye and 212.6 ± 123.3 µm in the

left eye. The mean post-ELD OCT-RNFL thickness was 149.4 ± 45 µm in the right eye and 151.4 ± 51.3 µm in the left eye. The mean duration of ELD was 8.7 ± 1.4 days (range, 7–10 days). In all children, the RNFL thickness had significantly decreased in both eyes after ELD ($p<0.05$).

Four patients (36.3%) required LPS surgery during the follow-up period. In two patients, although the RNFL thickness had decreased, an LPS was inserted just after ELD removal because the ICP was significantly high (Patients 8 and 9). In Patient 1, although the ICP did not decrease after ELD, LPS implantation was postponed because the RNFL thickness had significantly decreased in both eyes. However, the patient experienced a relapse with an increase in the RNFL thickness 2 weeks after ELD removal. Thus, an LPS was implanted. LPS implantation was also postponed in Patient 6 despite the absence of a significant decrease in RNFL thickness,

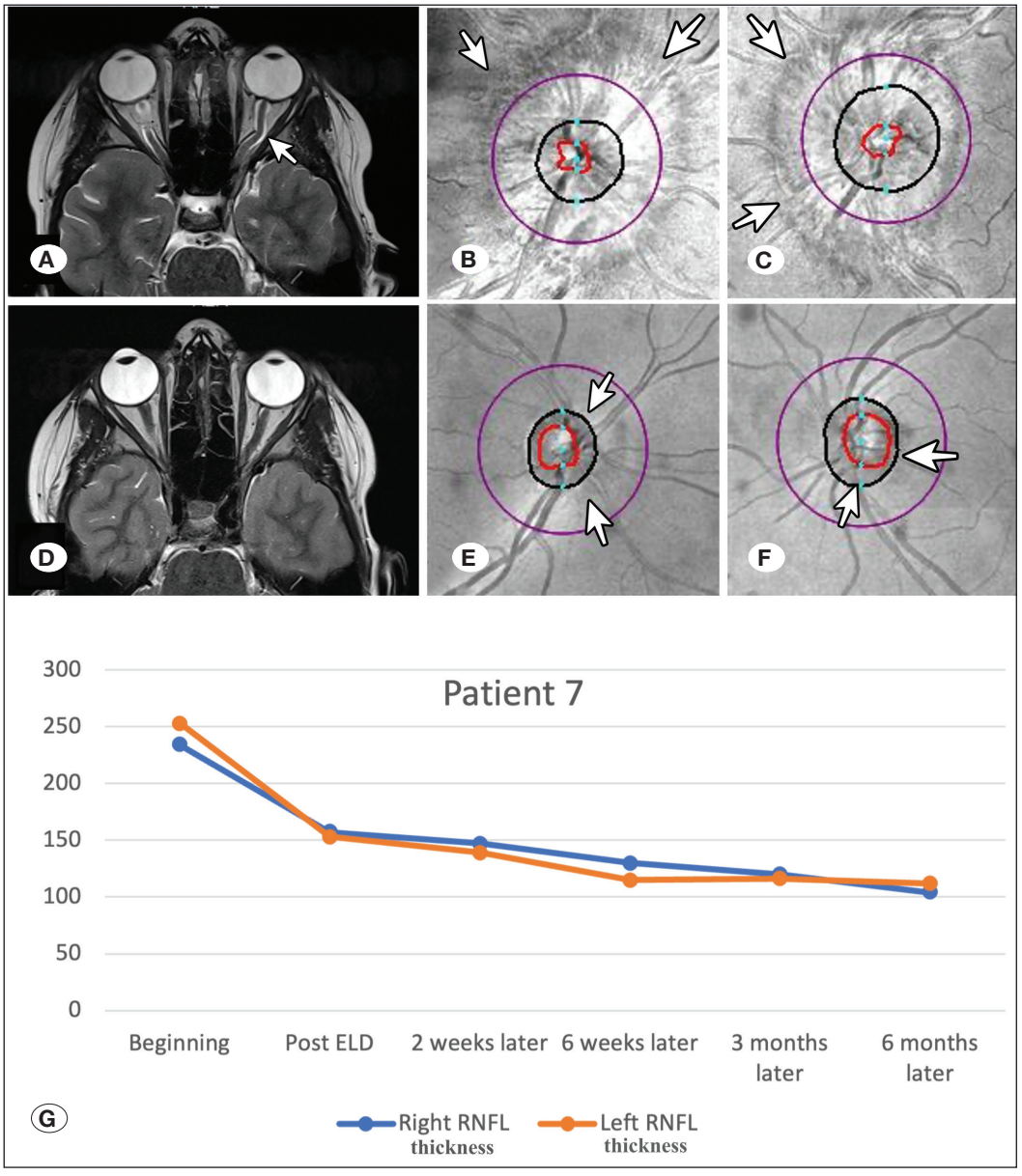

Figure 2: Images of representative Patient 7. **A)** T2-weighted axial MR image obtained before IIH treatment shows optic nerve sheath swelling with optic nerve kinking (arrow). OCT of the **(B)** right and **(C)** left eyes before IIH treatment show obvious swelling around the papillae (arrows). **D)** MRI obtained 6 months after IIH treatment (7 days of ELD + TPM, followed by TPM only) shows complete resolution of the optic nerve sheath swelling and optic nerve kinking. **E, F)** OCT obtained 6 months after the start of IIH treatment shows complete resolution of the swelling around the papilla. **G)** Graph showing the changes in OCT-RNFL thickness values within 6 months of treatment. There is a dramatic decrease in RNFL thickness at the end of the ELD treatment. The decrease in RNFL thickness continued during the follow-up period with oral treatment until physiological values were achieved. **MR:** Magnetic resonance, **IIH:** Idiopathic intracranial hypertension, **OCT:** Optical coherence tomography, **MRI:** Magnetic resonance imaging, **ELD:** External lumbar drainage, **TPM:** Topiramate, **RNFL:** Retinal nerve fiber layer thickness.

because there was a significant decrease in the ICP. However, IIH relapse was observed 4 weeks after ELD removal with an increase in ICP. Thus, she underwent LPS surgery. No relapse of IIH or complications such as infection and intracranial hypotension were reported during the follow-up period in any patient who had undergone LPS surgery. The remaining seven patients who did not undergo LPS surgery remained free of symptoms with a normal CSF pressure, improved ophthalmological findings, and no recurrence. Figure 2 shows the pre- and post-ELD cranial MR and OCT images of Patient 7 who did not undergo ELD. The graph indicates changes in the RNFL thickness with time. Graphs showing changes in RNFL thickness with time in other six patients (Patients 2, 3, 4, 5, 10, and 11) who did undergo ELD are depicted in Figure 3.

DISCUSSION

Although the exact pathophysiology of IIH is unknown, there are some theories that explain the possible mechanisms underlying the disease process (10,18,23-25). The most pronounced one is the collapse of the transverse sinuses, which leads to venous hypertension that decreases the CSF reabsorption and raises the ICP (10). This results in more pressure on the venous sinuses and completes the vicious cycle. However, the first event that triggers these cascade of events remains unknown. Some authors, while supporting this hypothesis, further proposed that development of venous hypertension before the closure of cranial sutures may cause hydrocephalus with dilated ventricles. However, development of venous hypertension after the closure of cranial sutures may cause IIH (18,19). There are several studies on the development of hydrocephalus in babies with achondroplasia and mucopolysaccharidosis in whom jugular foramen stenosis was determined as the cause for intracranial venous hypertension. This further supports the venous hypertension hypothesis (18,23,24).

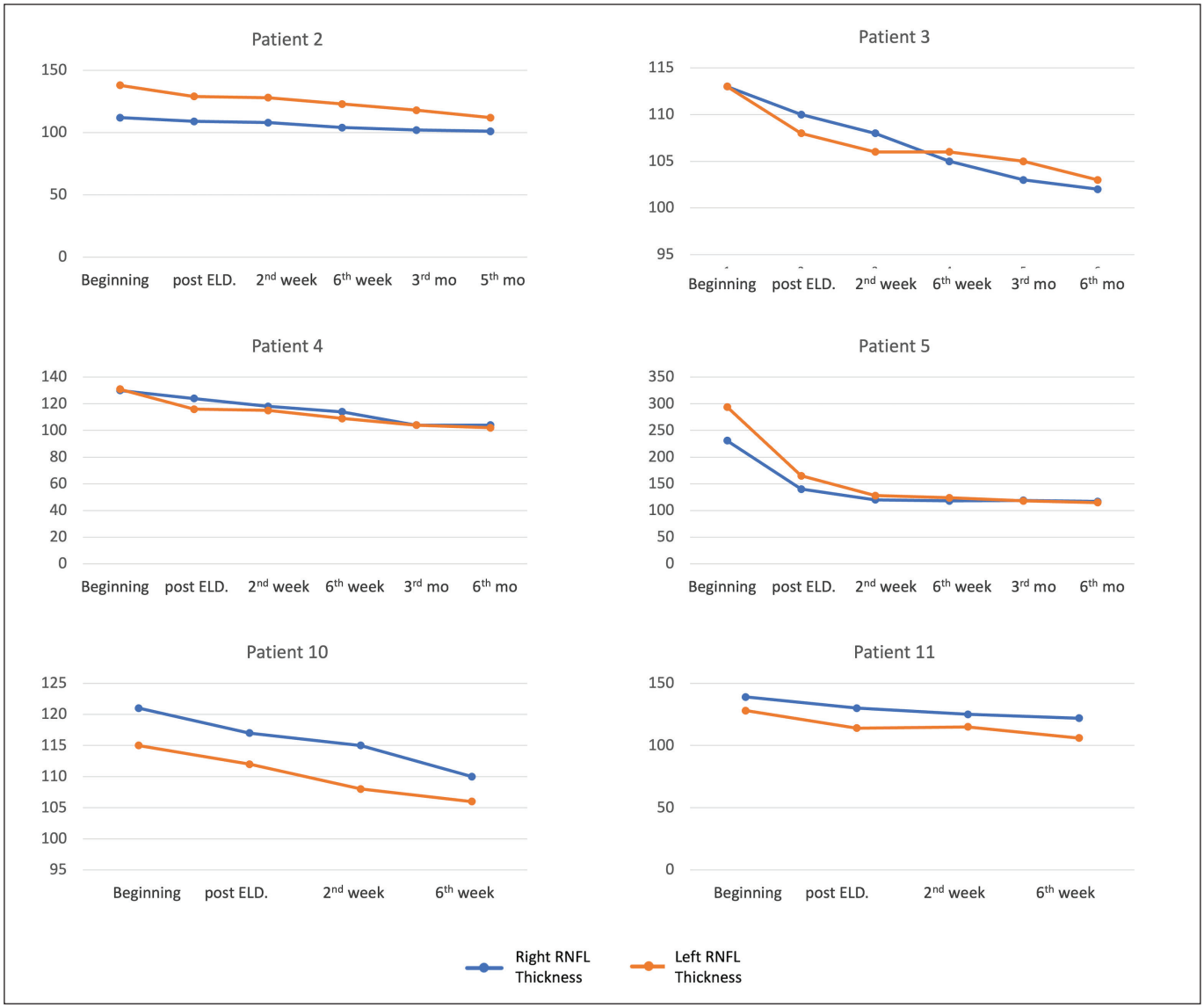

Figure 3: Graphs showing the changes in RNFL thickness over time in Patients 2, 3, 4, 5, 10, and 11.

The venous hypertension theory may also explain the higher incidence of IIH in obese patients. Obesity increases the intraabdominal pressure, which in turn increases the intrathoracic and right atrial pressure as well as the venous pressure within the superior vena cava, internal jugular vein, and, subsequently, intracranial venous sinuses that will impede CSF reabsorption (25).

Besides the more pronounced clinical presentations of IIH, such as headache and vomiting, the main concerns of most clinicians are the clinical or subclinical visual alterations due to optic nerve damage that is a result of chronically increased ICP (3,4). Overt papilledema can be easily diagnosed by an ophthalmologist. However, identification of subtle papilledema requires experience. Therefore, ophthalmological examination results for papilledema may exhibit interobserver variability. OCT can be used to measure RNFL thickness, and it yields objective and reproducible values. Furthermore, it diminishes interobserver variability, making it a practical tool for the diagnosis and follow-up of ophthalmological abnormalities, including papilledema (7,22).

All the patients in our study underwent an ophthalmological examination by the same ophthalmologist and an OCT at the time of admission, on the fifth day of ELD, at the end of ELD (7^{th}–10^{th} days), and on subsequent follow-ups. Similar to the findings of previous studies, we found that the OCT-RNFL values generally correlated with the ICP values during the follow-up (7,22).

Alleviation of symptoms and protection of vision are the two main treatment goals in IIH (4). The commonly used treatment options for IIH are oral medications, serial LP, and lifestyle modifications such as a specific weight-loss diet in overweight and obese patients. If these options fail, an LPS can be placed (3). Less commonly, transverse sinus stenting and ONSF may be attempted before LPS placement. However, they both are associated with considerable complication rates (3,7,11,13,15). The main rationale behind administering oral medications and serial LP is to decrease the intracranial pressure, allow the transverse sinuses to re-expand, and break the vicious cycle of ICP increase (7).

In nearly 85%–90% of the patients, oral medications are effective. However, the remaining patients generally require surgical intervention. The interval between oral acetazolamide use and resolution of ICP ranges between 40 and 90 days in adults (26). Schoeman reported the treatment results of combined acetazolamide and furosemide use, followed by the first LP in pediatric patients with IIH. They reported a decrease in the ICP values after 1 week of treatment, and normal values in all the patients within 6 weeks of treatment. Furthermore, remission of papilledema was observed in all the patients within 8 weeks of treatment (21). Although delayed remission of papilledema may be acceptable, normalization of ICP within 6 weeks after the start of oral medication may be perilous in terms of visual protection in pediatric patients with fulminant IIH.

Serial LPs are uncomfortable and generally not recommended because of complications such as infection, bleeding, and the inclusion of dermoid cysts. Furthermore, while some authors question its therapeutic benefit, others believe that serial LPs are not useful in IIH (2,4,7,12). The therapeutic use of serial LPs is based on the theory of increased ICP and compressed intracranial venous sinuses. However, considering the CSF production rate, a daily withdrawal of a few milliliters of CSF will not interrupt the vicious cycle because it will be reproduced within minutes (29).

Because oral medications require more time to act and serial LPs are ineffective, ELD was considered to continuously drain more CSF under sterile conditions and monitor the ICP. Continuous ICP monitoring helps adjust the CSF drainage rate. None of the patients in our study developed hemorrhagic, infectious, or neurological complications, and all patients demonstrated significant improvement in the ICP and OCT-RNFL thickness values. Including the patients who developed relapses, none of the participants experienced worsening visual functions. After retrospectively evaluating the results of four patients who underwent LPS surgery, we propose that discordant changes in ICP and RNFL thickness values may be an indication for LPS surgery.

Studies on ELD use in pediatric IIH are limited. Furthermore, in these studies, ELD was only used in patients in whom oral medications failed, which was associated with worsening papilledema and/or visual functions (7,10,14). Compared with previous studies that used oral medications alone, we used a combination of oral medications and ELD to enhance resolution of papilledema and ICP reduction (21,26). Our findings suggests that early intervention with ELD, rather than waiting for medication failure, may better preserve visual function in pediatric patients. Moreover, the use of OCT allowed us to objectively monitor the resolution of papilledema, which may have been missed with fundus examination alone

The results of our study demonstrate that the combination of ELD and oral medication was effective in achieving a rapid reduction in the ICP and RNFL thickness. This is particularly significant because prolonged elevated ICP can cause irreversible optic nerve damage. In our cohort, we observed a marked decrease in both ICP and RNFL thickness after ELD, which persisted over the short-term follow-up. This finding aligns with those of previous studies, which also demonstrated the effectiveness of ELD in lowering ICP and protecting against optic nerve damage in patients with IIH (7).

The difference in the follow-up duration between patients who required an LPS and those who did not is notable. Patients without LPS tended to have a shorter follow-up period, because their condition stabilized with the combined ELD and medication approach. This may indicate a faster remission. However, the short follow-up period limits our ability to assess the long-term outcomes such as recurrence of papilledema or elevated ICP. Patients 10 and 11, with notably shorter follow-up periods of 2 months, did not experience a recurrence. However, long-term observation is required to verify the sustained remission. The short follow-up period and considerably small sample size are limitations of the study.

CONCLUSION

Proactive ELD use is an effective method to achieve a rapid decrease in ICP and RNFL thickness with no major complications. Thus, we propose combining ELD with oral medications as a first-line treatment option in pediatric patients with IIH in whom ICP values are more than 300 mmH_2O.

Declarations

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Availability of data and materials: The datasets generated and/or analyzed during the current study are available from the corresponding author by reasonable request.

Disclosure: The authors declare no competing interests.

AUTHORSHIP CONTRIBUTION

Study conception and design: BT, MMO

Data collection: BT, UI, MAT

Analysis and interpretation of results: BT

Draft manuscript preparation: BT

Critical revision of the article: MMO

All authors (BT, MAT, UI, MMO) reviewed the results and approved the final version of the manuscript.

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