(02)

The importance of visual field testing in patients with papilledema and idiopathic intracranial hypertension

Znaczenie badania pola widzenia u pacjentów z obrzękiem tarczy nerwu wzrokowego i idiopatycznym nadciśnieniem śródczaszkowym

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Abstract:
Aim: To determine the role of visual field testing in the monitoring of patients with idiopathic intracranial hypertension.
Material and methods: 48 patients with newly diagnosed idiopathic intracranial hypertension were enrolled. Routine neurologic and ophthalmic examinations as well as automated perimetry were performed in all patients at baseline and 4–6 months later. Based on baseline findings, the subjects were referred for further treatment which included reduction of body weight, sodium-restricted diet, oral acetazolamide or other diuretic agent, corticosteroids and, in three cases, also optic nerve sheath decompression.
Results: 39 patients (81%) presented with visual field defects on baseline perimetry. The most commonly found visual field defects included blind spot enlargement, usually with concomitant partial peripheral rim (29 eyes, 38.7%), generalized constriction (16 eyes, 21.3%) and nerve fiber bundle defect with or without concomitant blind spot enlargement (15 eyes, 20%). In 28 patients (58%), the findings of follow-up perimetry differed from baseline. Visual field defects were found in 3 persons with normal visual field at baseline. Up 60% of treated patients with abnormal baseline perimetry showed an improvement in visual field parameters, whereas 86% of untreated patients had no change in visual field parameters whatsoever.
Conclusions: Treatment of patients with idiopathic intracranial hypertension contributes to the improvement of visual field parameters. Ophthalmic examination with perimetry is a critical component of baseline status evaluation, treatment recommendations and monitoring treatment outcomes in patients with idiopathic intracranial hypertension.
Key words: papilledema, visual field, idiopathic intracranial hypertension.
Abstrakt:
Cel: podkreślenie roli badania pola widzenia w monitorowaniu pacjentów z idiopatycznym nadciśnieniem śródczaszkowym.
Materiał i metody: do badań włączono 48 pacjentów ze świeżo rozpoznanym idiopatycznym nadciśnieniem śródczaszkowym. U wszystkich chorych wykonano wstępne rutynowe badania neurologiczne i okulistyczne, poszerzone o automatyczną perymetrię, oraz badanie kontrolne po 4–6 miesiącach leczenia. Na podstawie pierwszego badania kwalifikowano pacjentów do leczenia, które obejmowało obniżenie masy ciała, stosowanie diety z niską zawartością soli, doustną podaż acetazolamidu lub innego diuretyku, podawanie steroidów oraz dekompresję osłonek nerwu wzrokowego, którą wykonano w 3 przypadkach.
Wyniki: ubytki w polu widzenia w wyjściowej perymetrii prezentowało 39 chorych (81%). Najczęściej stwierdzanymi defektami pola widzenia były: poszerzenie plamy ślepej, zazwyczaj skojarzone z częściowym ubytkiem obwodowego pola (29 oczu, 38,7%), uogólnione zawężenie obwodu (16 oczu, 21,3%) oraz ubytek pęczkowy włókien nerwowych z poszerzeniem plamy ślepej lub bez jej poszerzenia (15 oczu, 20%). U 28 pacjentów (58%) wynik kontrolnej perymetrii różnił się od wyniku wyjściowego. U 3 osób, u których wynik pierwszego badania perymetrycznego był prawidłowy, stwierdzono ubytki w polu widzenia. W grupie pacjentów z nieprawidłowym wynikiem pierwszej perymetrii poprawę parametrów pola widzenia zauważono u 60% leczonych, a brak poprawy, a nawet pogorszenie, u 86% nieleczonych.
Wnioski: u pacjentów z idiopatycznym nadciśnieniem śródczaszkowym leczenie poprawia parametry pola widzenia. Badanie okulistyczne, w tym perymetria, ma kluczowe znaczenie w procesie kwalifikacji pacjentów do leczenia, ocenie leczenia oraz monitorowaniu jego wyników u pacjentów z idiopatycznym nadciśnieniem śródczaszkowym.
Słowa kluczowe: obrzęk tarczy nerwu wzrokowego, pole widzenia, idiopatyczne nadciśnienie śródczaszkowe.

Introduction

The term "idiopathic intracranial hypertension" (IIH) is referred to as an increase in intracranial pressure (ICP) of undetermined etiology (1). IIH is diagnosed whenever despite an evident clinical manifestation of elevated ICP, brain imaging reveals no underlying pathology, the patient does not present with focal neurological symptoms, and spinal tap confirms elevated pressure of otherwise biochemically and cytologically normal cerebrospinal fluid.

The vast majority of patients with IIH present on funduscopy with papilledema, usually binocular, although not necessarily symmetrical. Elevated pressure of cerebrospinal fluid in the subarachnoid space between the optic nerve and its sheath contributes to axonal compression, impairment of axoplasmic flow and resultant axoplasmic stasis in the optic disc. Eventually, papilledema develops within the first five days following the ICP elevation. Initially, the signs of edema can be seen in the superior and inferior part of the disc, gradually involving its nasal, and eventually also temporal part. Axonal compression leads to visual loss, which manifests as visual field defects. Perimetric abnormalities are found in 75–95% of patients with papilledema (2–5).

Due to the absence of other objective neurological symptoms, detailed ophthalmic examination, including perimetry, becomes a vital diagnostic option in patients with suspected IIH. Consequently, detection of papilledema and assessment of visual function are essential components of IIH management. The aim of this paper is to demonstrate the role of visual field testing in the monitoring of IIH patients.

Material and methods

48 patients (43 women and 5 men), aged between 28 and 59 years (mean age of 46 ± 8.2 years) with papilledema due to newly diagnosed IIH satisfying the modified Dandy criteria were enrolled (6). All subjects have been treated or consulted as inpatients or outpatients at the Department of Ophthalmology, Medical University of Bialystok. The only exclusion criterion was the presence of another ocular or neurologic pathology.

All patients underwent a routine neurologic assessment, as well as ophthalmic assessment with perimetry. Based on the neurologic and ocular findings, they were referred for further treatment, which included weight loss program, sodium-restricted diet, oral acetazolamide or other diuretic agent, corticosteroids and, in three cases, also optic nerve sheath decompression. A total of 25 patients received the treatment; others were either found ineligible for treatment, did not give their informed consent for the intervention, or discontinued treatment at early stages. All subjects were re-examined by the ophthalmologist 4–6 months later. The outcomes were analyzed separately for three groups: 1. subjects left untreated due to lack of visual field defects on baseline perimetry, 2. subjects, who received treatment, due to presence of visual field defects, and 3. subjects, who did not receive treatment, despite abnormal findings on perimetry.

Visual fields were assessed with automated perimetry (Medmont Model M700) using full threshold strategy. A standardized grid of 164 static targets within the central 50 degrees of the visual field was applied with decreased stimulus intensity in steps of 3 dB, until threshold sensitivity was reached. Appropriate correction for near vision was included. Reliability criteria were established in line with the recommended standards, i.e. less than 20% fixation losses, false-negative error and false-positive error rates below 33% each. If the reliability criteria were not satisfied, perimetry was repeated during the second session. Visual field defect patterns and binocular characteristics were evaluated.

Visual field defects were categorized according to their predominant pattern. The result of perimetry was classified as normal visual field, blind spot enlargement, nerve fiber bundle defect (pericentral, partial arcuate, arcuate), altitudinal defect, partial peripheral rim, generalized constriction and diffuse visual field loss. Visual field in patients with small superior visual field defects was classified as normal whenever the lid artifact was suspected.

The study protocol was approved by the University Institutional Review Board (according to the guidelines of the Helsinki Declaration), and all patients gave their written consent for the use of their clinical material in this publication.

Results

A total of 39 patients (81%) presented with visual field defects on baseline perimetry. Overall, visual field defects were found in 75 eyes, bilaterally in 36 patients and unilaterally in 3. In 8 patients with binocular lesions, the defects found in the right and left eye were essentially identical. In other 18 patients, binocular lesions differed in size and severity, although belonging to the same category of visual field defects. Finally, 10 patients showed significant differences in the location of scotomas between the right and left eye.

The list of the most commonly found visual field defects included blind spot enlargement, usually with concomitant partial peripheral rim (29 eyes, 38.7%), followed by generalized constriction (16 eyes, 21.3%) and nerve fiber bundle defect with/without concomitant blind spot enlargement (15 eyes, 20%). The least common defects were diffuse visual field loss (6 eyes, 8%), altitudinal defect (5 eyes, 6.7%), and only partial peripheral rim (4 eyes, 5.3%).

In 28 patients (58%), the result of follow up perimetry performed 4–6 months later was different than at baseline. Visual field defects manifesting as paracentral scotomas with concomitant blind spot enlargement were found in 3 persons with normal baseline perimetry. Up to 60% of treated patients with abnormal baseline perimetry showed an improvement in visual field parameters, whereas 86% of untreated patients showed no change in visual field parameters whatsoever. The results of follow up perimetry are summarized in Table I.

	Group 1/ Grupa 1.	Group 2/ Grupa 2.	Group 3/ Grupa 3.
Worsening/ Pogorszenie	3	2	6
No change/ Bez zmian	6	8	6
Improvement/ Poprawa	-	15	2
48 patients/ 48 pacjentów	**9**	**25**	**14**

Tab. I. Changes of visual field parameters in patients with papilledema and idiopathic intracranial hypertension, documented on follow up perimetry after 4–6 months of treatment.

Tab. I. Zmiany w parametrach kontrolnego badania pola widzenia wykonanego po 4–6 miesiącach leczenia u pacjentów z obrzękiem tarczy nerwu wzrokowego i idiopatycznym nadciśnieniem śródczaszkowym.

Figures 1 to 10 document the improvement in the visual field parameters of 5 patients with IIH. Patients #1, 3, 4 and 5 were treated with acetazolamide and sodium-restricted diet, whereas Patient #2 underwent endoscopic procedure of optic nerve sheath decompression.

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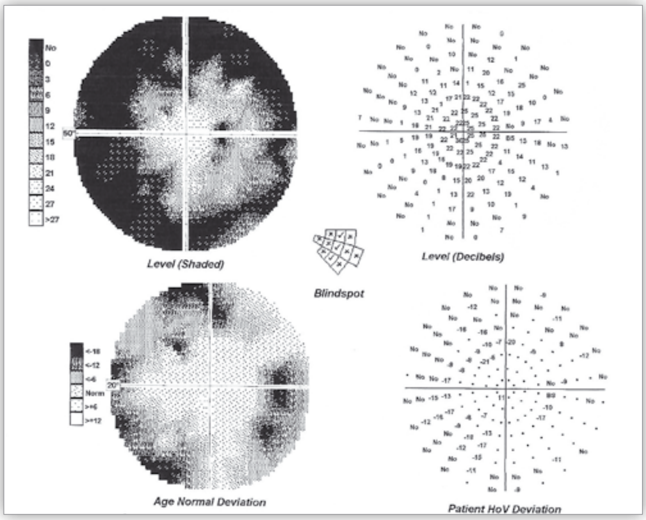

Fig. 1. Automated computed perimetry of the right eye in Patient #1 (baseline assessment).
Ryc. 1. Automatyczna perymetria komputerowa prawego oka u pacjenta nr 1 (badanie wstępne).

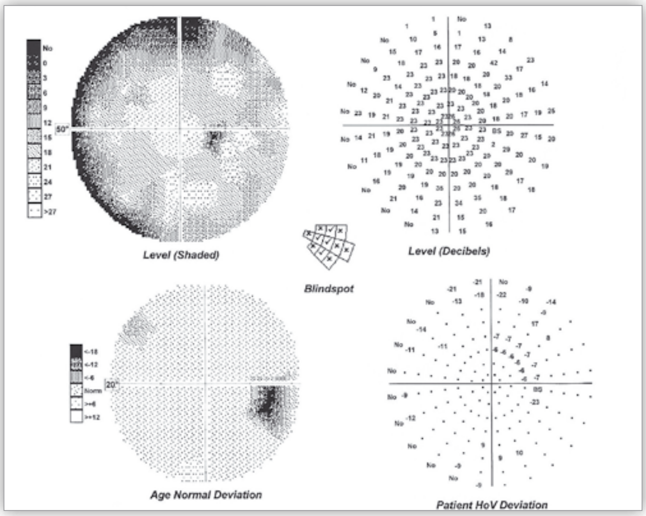

Fig. 2. Automated computed perimetry of the right eye of Patient #1 (follow-up assessment).
Ryc. 2. Automatyczna perymetria komputerowa prawego oka u pacjenta nr 1 (badanie kontrolne).

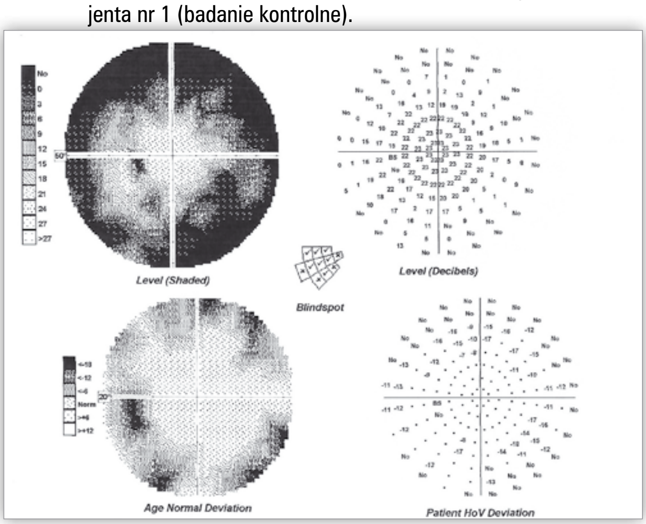

Fig. 3. Automated computed perimetry of the left eye of Patient #2 (baseline assessment).
Ryc. 3. Automatyczna perymetria komputerowa lewego oka u pacjenta nr 2 (badanie wstępne).

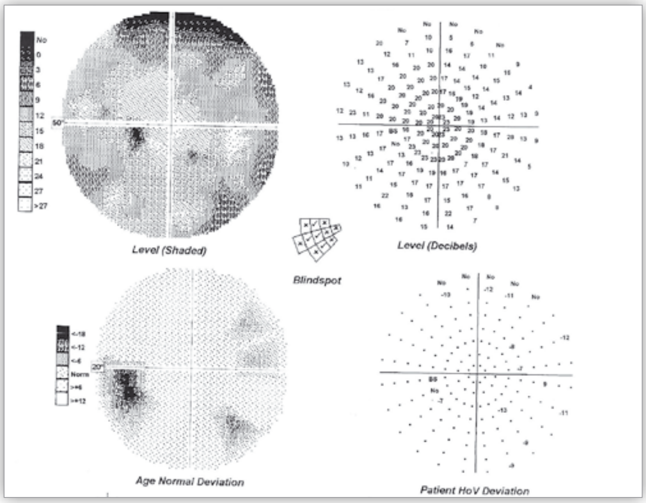

Fig. 4. Automated computed perimetry of the left eye of Patient #2 (follow-up assessment).
Ryc. 4. Automatyczna perymetria komputerowa lewego oka u pacjenta nr 2 (badanie kontrolne).

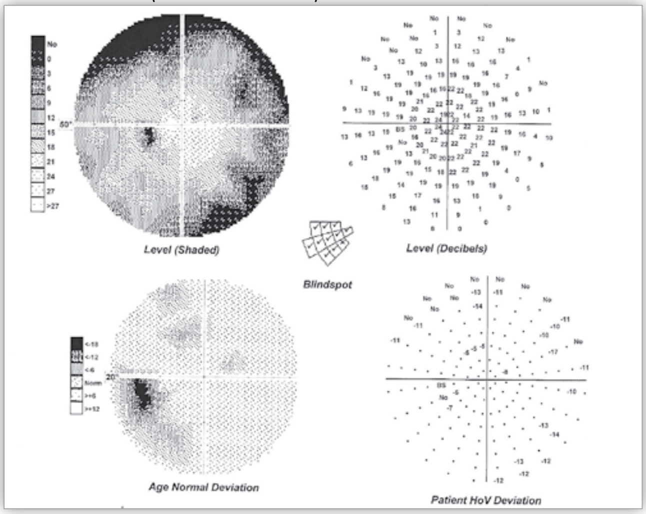

Fig. 5. Automated computed perimetry of the left eye of Patient #3 (baseline assessment).
Ryc. 5. Automatyczna perymetria komputerowa lewego oka u pacjenta nr 3 (badanie wstępne).

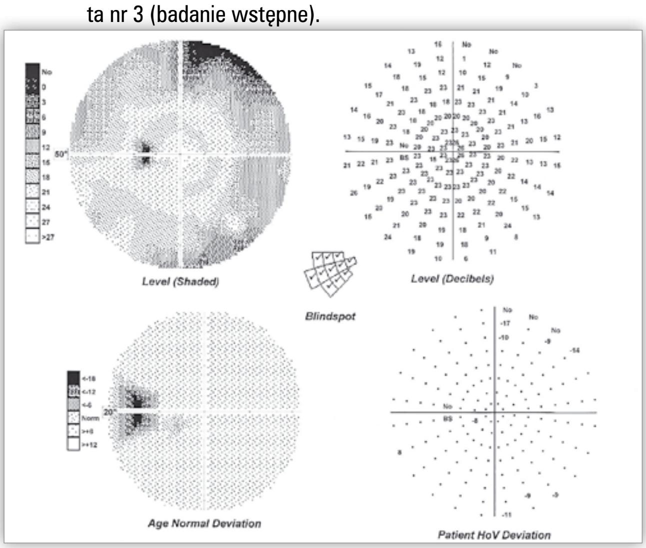

Fig. 6. Automated computed perimetry of the left eye of Patient #3 (follow-up assessment).
Ryc. 6. Automatyczna perymetria komputerowa lewego oka u pacjenta nr 3 (badanie kontrolne).

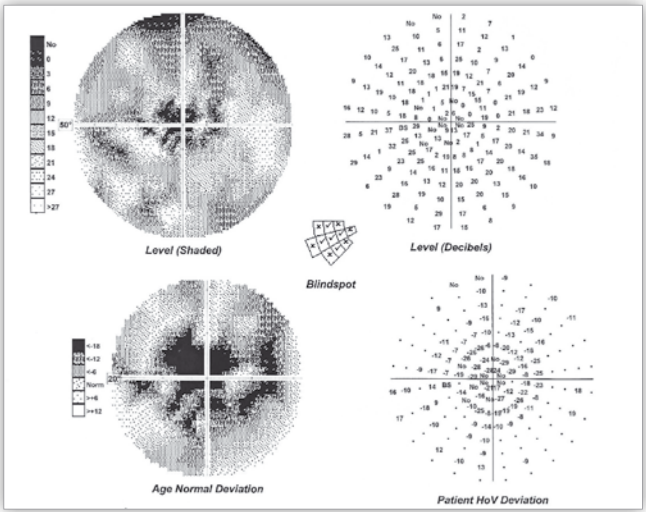

Fig. 7. Automated computed perimetry of the left eye of Patient #4 (baseline assessment).
Ryc. 7. Automatyczna perymetria komputerowa lewego oka u pacjenta nr 4 (badanie wstępne).

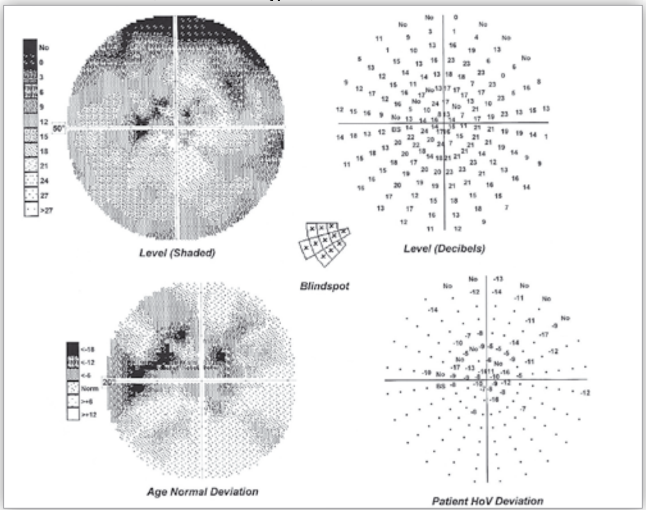

Fig. 8. Automated computed perimetry of the left eye of Patient #4 (follow-up assessment).
Ryc. 8. Automatyczna perymetria komputerowa lewego oka u pacjenta nr 4 (badanie kontrolne).

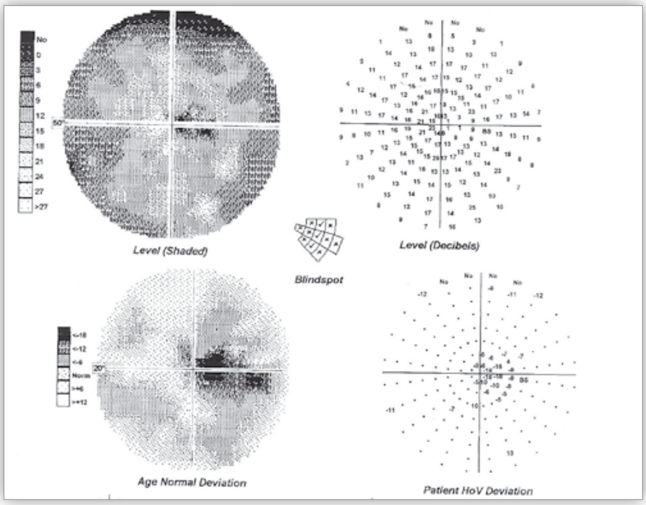

Fig. 9. Automated computed perimetry of the right eye of Patient #5 (baseline assessment).
Ryc. 9. Automatyczna perymetria komputerowa prawego oka u pacjenta nr 5 (badanie wstępne).

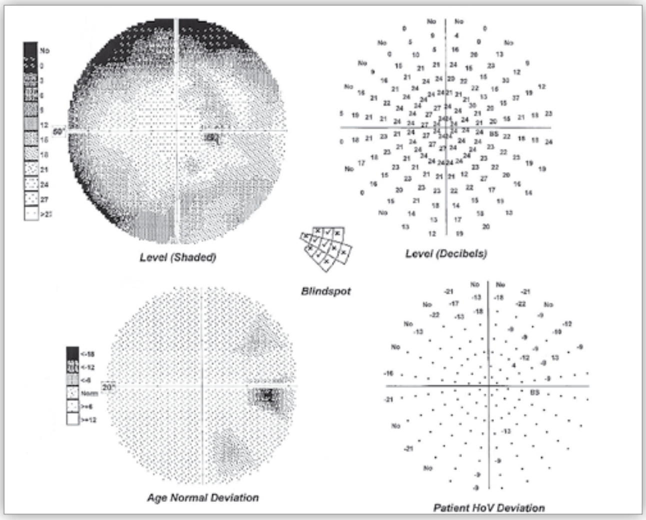

Fig. 10. Automated computed perimetry of the right eye of Patient #5 (follow-up assessment).
Ryc. 10. Automatyczna perymetria komputerowa prawego oka u pacjenta nr 5 (badanie kontrolne).

Discussion

In this study, up to 81% of patients with papilledema and IIH presented with visual field defects on baseline perimetry. The principal underlying mechanism of visual loss in these patients was probably a decrease in axonal flow or complete disruption thereof, resulting from an increase in cerebrospinal fluid pressure, transmitted down the optic nerve sheath. Axoplasmic flow stasis contributes to intraneuronal ischemia of the optic disc (7, 8). Other, less common causes of visual field defects include fluid tracking from the optic disc to the fovea with resultant neurosensory retinal detachment, or hyperopic shifts associated with optic nerve sheath-related globe flattening and elevation of peripapillary retina (9).

The most common types of visual field defects found in our patients with IIH were blind spot enlargement, nerve fiber bundle-like defects and generalized constriction. A large body of evidence suggests that optic disc is the main site of damage in papilledema. The defects associated with papilledema are known to closely resemble those observed in glaucomatous neuropathy (10).

The most common types of visual field defects present in the series of 50 patients with IIH examined by Wall and George (11) were blind spot enlargement, generalized constriction and loss of nasal (especially inferonasal) visual field. In turn, the most prevalent hemifield abnormality found during the baseline examination of patients participating in the Intracranial Hypertension Treatment Trial (IIHTT) was localized nerve fiber bundle-like defect with or without a concomitant blind spot enlargement (71.5%) (9). The same study demonstrated that visual field defects related to IIH are more frequent in the inferior hemisphere and in the left eye.

The role of perimetry in patients with IIH is not limited to the source of information about actual degree of visual loss. Visual field testing is particularly useful in monitoring progress of IIH. Abnormal perimetric findings constitute a firm indication for causal treatment of this condition (12), and appropriate treatment eligibility assessment is crucial considering its various

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potential side effects. Currently, a standard treatment for IIH is oral acetazolamide, combined with sodium-restricted diet and weight loss. A few published studies endorse such approach as highly effective and likely to improve visual field parameters in more than 50% of cases (13, 14). The current findings are consistent with this data, since up to 60% of patients treated for IIH showed an improvement of visual function at the end of 4–6-month follow-up. The improvement was particularly evident after endoscopic optic nerve sheath decompression, but only few patients opted for this procedure due to associated risks of vision loss.

According to Wall (4), approximately one third of patients with IIH and visual field defects may be unaware of the latter if localized within the peripheral visual field. This is an additional argument for a comprehensive ocular assessment in subjects with established IIH and individuals reporting frequent and severe headaches, a likely manifestation of this condition. As already mentioned, visual field defects are reversible if detected and treated early enough.

In conclusion, visual field testing plays a key role in monitoring of patients with papilledema and IIH. Since subjects with IIH typically present with normal results of other tests, including neurologic exam and diagnostic imaging, perimetry is the only objective non-invasive procedure to document the dynamics of ICP-related changes. Visual field testing is particularly important in the monitoring of therapeutic outcomes in patients treated for IIH. Since visual loss is reversible with early causal treatment, all patients with IIH should undergo regular ocular assessment including perimetry.

Conclusions

1. A large proportion of patients with papilledema and IIH present with visual field defects.
2. Blind spot enlargement with partial peripheral rim, general constriction and nerve fiber bundle-like defects are typical features of optic nerve damage associated with IIH.
3. Anti-IIH treatment contributes to the improvement of visual field parameters.
4. Ophthalmic examination with perimetry is a critical component of baseline status evaluation, treatment recommendations and monitoring treatment outcomes in patients with IIH.

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