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Idiopathic intracranial hypertension: Proposal of a stratification strategy for monitoring risk of disease progression

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ABSTRACT

Objectives: A general consensus based on a multidisciplinary perspective involving an international panel was recently developed for management of patients with idiopathic intracranial hypertension (IIH). In this paper, the authors sought to develop further on the aspect of this consensus that concerns monitoring progression of the disease.

Patients and methods: A systematic literature review of previous publications on monitoring disease progression in IIH and a meta-analysis to examine efficacy of method of monitoring employed in each study. The authors present a brief descriptive analysis of challenges with monitoring progression of the disease and propose a risk stratification to aid monitoring.

Results: Of a total of 382 publications identified from the literature search, only 8 studies (144 patients) satisfied inclusion criteria and were included for analysis. Among these, 3 were based on ICP monitoring while the remaining 5 focused on ophthamological evaluation. Interestingly, there were neither any studies on monitoring with progression of clinical features nor any study on monitoring with symptomatology associated with IIH among the selected studies.

Conclusion: There is a paucity of studies in the literature on methods of monitoring disease progression in IIH. Though close attention to adequate evaluation and proper care of patients with IIH remains the key in managing this problem, this proposed risk stratification will be an objective tool and useful guide to better monitor these patients according to their extent of risk from the disease and possibly for planning treatment and intervention.

1. Introduction

Idiopathic intracranial hypertension, IIH (also known as benign intracranial hypertension and as pseudotumour cerebri) is the clinical syndrome of raised intracranial pressure, but in the absence of any space-occupying lesions or vascular lesions, intracranial infections, hydrocephalus or hypertensive encephalopathy and for which no causative factor can be identified [1–5]. Hence, it is a diagnosis of exclusion [1,2,6]. It was first described as 'serous meningitis' by Quinke in 1893 and was later named by Foley in 1955 [6,7]. Various criteria previously proposed to aid in its diagnosis include Dandy's criteria and Friedman's criteria [1,5,7,8]. It tends to commonly occur in women of child bearing age who are obese [1–3,7–9]. It is a well documented health problem with incidence ranging from 0.03 to 2.2 per 100,000 based on various reports and as much as 19–21 per 100,000 has been recorded among women of child bearing age though well reported in men [4], and with documented female to male ratios widely ranging from 2:1 to 15:1 [1,2,4,7]. However, it is not limited to adults [6,10–12]. Even children as young as four months old have been reported to have the problem [6,13,14].

Currently, the universally accepted aim of its management is primarily to halt or prevent visual loss [1,6,15–17]. This may occur early

Abbreviations: PRISMA, preferred reporting items for systematic reviews and meta-analysis; IIHWOP, idiopathic intracranial hypertension without papilledema; CSF, cerebrospinal fluid; BMI, body mass index; IIH, idiopathic intracranial hypertension; ID, identity; LP, lumbar puncture

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or late in the course of the condition [16]. Time of its diagnosis within 6 months of its onset has been identified as the only factor significantly associated with visual improvement [18]. However, much of what is currently known and generally accepted regarding its management is controversial [1]. Being a diagnosis of exclusion, variations in its clinical presentation may make diagnosing it somewhat challenging [3].

Recently, statement guidelines based on general consensus from a multidisciplinary perspective by an international panel of experts cutting across various professional bodies were developed for the investigation and management of patients with IIH [65]. In this article, we sought to develop further on the aspect of the current guidelines on monitoring by systematic review of the literature for studies focusing on monitoring of disease progression in IIH with the aim of identifying clinical parameters which may be useful for monitoring progression of the disease. We carry out a meta-analysis to evaluate effectiveness of the methods of monitoring employed in the selected studies, and then suggest a risk stratification as an aid towards monitoring progression of the disease (particularly with emphasis on risk of visual loss).

2. Materials and methods

The Preferred Reporting Items for Systematic reviews and Meta-analysis (PRISMA) was utilized for this review (Fig. 1). An initial systematic search of peer-reviewed literature and previous studies on IIH was performed using the MeSH term “benign intracranial hypertension OR idiopathic intracranial hypertension AND (monitoring)” in PubMed database and Google search engine and also with the key phrases “benign intracranial hypertension and monitoring with symptoms”, “intracranial pressure monitoring in pseudotumour cerebri”, “monitoring idiopathic intracranial hypertension with neurologic deficits”, and “monitoring for intracranial pressure with visual loss”. The search was limited to only articles published in English language (Table1). The search was limited to only published studies; results of unpublished studies or abstracts of studies whose full articles were not available were also excluded (Table1). The reference section in these articles were also reviewed for any further relevant material. All selected papers from the search were each assigned a Study ID (identity) number. A meta-analysis of the selected articles was performed using Wilson Macros for Meta-analysis in SPSS software to examine how effective the method of monitoring employed in each study is. Statistical heterogeneity among the selected studies was calculated using Cochran I^2 and Q statistics [19]. Results were reported with their 95% confidence interval (CI). The significance level was $p < 0.05$. Random or fixed-effect meta-analysis was performed, depending on heterogeneity of the data. Where statistically significant heterogeneity was found, the random effects model was used to combine results. Otherwise, the fixed effects model was used. Subgroup analyses were performed based on method of monitoring involved. ROBINS-I (Risk Of Bias In Non-Randomised Studies – of Interventions) tool was used to evaluate for bias among the selected studies by using its “signaling questions” to judge risk of bias in each domain [20], and Forest plot analysis carried out for the selected studies. All analyses were based on previously published studies and hence, no ethical approval or patient consent was required. In the Discussion section, we present a brief descriptive analysis of challenges with monitoring progression of the disease using 1.) its signs and symptomatology, 2.) intracranial pressure changes, 3.) its effect on vision as well as 4.) investigations and then propose a risk stratification strategy.

3. Results

3.1. Baseline characteristics

A total of 382 publications were identified from the electronic database search. After applying the exclusion criteria, 50 references remained for full text evaluation (Fig. 1). Manual reference list searches did not yield any additional studies. After final application of criteria, there were 8 studies (144 patients) included for qualitative and quantitative analysis in this study (Fig. 1). Among these 8 selected publications, 3 were based on ICP monitoring (Study ID 74, 319 and 313)

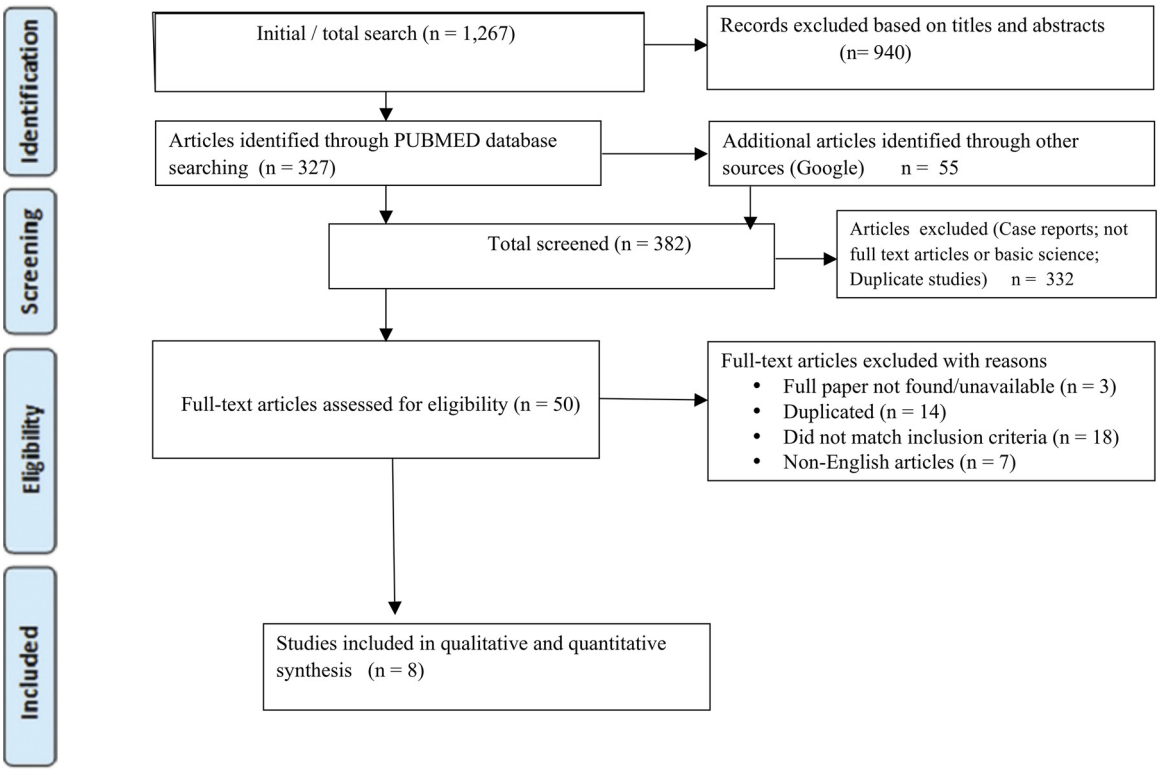

Fig. 1. PRISMA flow chart of systematic review and study selection process. (PRISMA, Preferred Reporting Items for Systematic Reviews and Meta-Analyses.).

Table 1
Summary of selection criteria KEY: IIH – Idiopathic intracranial hypertension.

INCLUSION CRITERIA	EXCLUSION CRITERIA
Only articles involving methods of monitoring progression of IIH	Studies involving monitoring for other causes of hypertension apart from IIH
Studies involving only humans	Articles on only methods of assessment of IIH, but not monitoring its progression
Observational study (Case control or Cohort)	Unpublished studies, Case reports, case series, Reviews, editorials on IIH
Only articles in English language	Articles in other languages

Table 2
Baseline data for selected studies. KEY: SDOCT – 3 dimensional spectral domain optical coherence tomography, OCT – optical coherence tomography, ICP – intracranial pressure.

Year of publication	Method(s) of monitoring disease progression evaluated	Study population (*n*)	Duration of monitoring	Authors
2011	Continuous ICP monitoring with intraparenchymal Codman ICP Monitoring System	10	short-term	Warden et al.
2007	Laser scanning tomography (LST) measurement of optic disc for papilledema	24	12 months	Heckmann et al.
1988	Spatial contrast sensitivity to detect visual loss	20	1 – 60 months	Bulens et al.
1978	Long-term ICP monitoring by implanted pressure-sensing capsule.	4	10 - 22 months	Gucer et al.
2017	Analysis of retinal morphology and optic nerve head morphology with SDOCT	21	19.2 months	Albrecht et al.
2009	A. Visual field assessment B. Measurement of peripapillary retinal nerve fiber layer (RNFL) thickness with OCT	22	12 months	Rebolleda et al.
1998	A. Visual field assessment with Humphrey and Goldmann perimeters, B. Visual acuity and contrast sensitivity.	35	3 years	Rowe et al.
1979	Long-term ICP monitoring with implanted Hittman-Meyer ICP sensor	8	14 months	Cooper et al.

while the remaining 5 focused on serial ophthamological evaluation (Table 2) in form of fine high-resolution neuroimaging (Study ID 102, 250, 6, 91 and 138). Of the 5 studies on serial ophthamological evaluation category, 2 studies (Study ID 91 and 138) involved 2 different methods each and hence were subcategorized into 91 A, 91B and 138 A, 138B respectively. Interestingly, there were no studies on monitoring with progression of clinical features associated with IIH among the selected studies.

However, in evaluating laser scanning tomography measurements of the extent of papilledema as a monitoring parameter, only Study ID 102 compared measurements with clinical parameters from among all 8 selected studies.

The overall mean effect size for all the 8 studies by the fixed effect model was 0.397 (95% CI = 0.25 – 0.55; Z = 5.19) and the weighted standard deviation was 0.440. For subgroup analyses which was based on method of monitoring involved in the selected studies, the mean effect size for studies utilizing ICP for monitoring was 1.308 (95%CI = 0.64 – 0.96; Z = 4.72), while for studies based on optic disc / visual assessment methods of monitoring, it was 0.322 (95%CI = 0.16 – 0.44; Z = 4.04). Statistical heterogeneity was significant among the selected studies (I^2 = 72.8%; Q = 33.063; p = 0.0001) and hence, the random effects model was used with mean effect size of 0.487 (95%CI = 0.18 – 0.80; Z = 3.09; p = 0.0020). For subgroup analyses, statistical heterogeneity was similarly significant among studies utilizing ICP monitoring (I^2 = 99.3%; Q = 1.0035; p = 0.6055) but not significant among studies utilizing optic disc / visual assessment methods (I^2 = 65.7% ; Q = 20.379; p = 0.0024). There was however no statistically significant intergroup difference between both subgroups (Q = 11.681; p = 0.166). Hence, method of monitoring for progression of IIH as the grouping variable did not account for the significant variability in effect sizes. Fig. 2 shows the Forest plot of the selected studies while Table 3 summarizes their risk-of-bias assessment.

4. Discussion

There is a paucity of studies on methods of monitoring disease progression in IIH as evidenced by only a few publications found from our literature search. Its rare nature may possibly account as the reason for this finding. The lack of studies on the issue as well as the varied methods of monitoring employed in the few selected studies are possibly the key factors which may account for the marked statistical heterogeneity of about 72.8% as observed. Results of analysis for these few selected studies in two subgroups suggest neither methods involving ICP nor methods utilizing optic disc / visual assessment methods to be a more superior or more sensitive method of monitoring disease progression. This implies that none of the methods evaluated in the selected publications may be a more effective method of monitoring progression of the disease compared to the others. The few number of available studies in the literature may also account for this. In view of the current guidelines on management of IIH [65], we examine in detail other methods that could possibly be utilized and their pitfalls, to arrive at our proposed risk stratification strategy.

4.1. Monitoring with Signs and symptomatology

Regarding symptomatology and signs related to rise in intracranial pressure as a result of IIH, these may include headache, nausea, vomiting, pulsatile tinnitus, transient visual obscurations, papilledema, CSF rhinorrhea and even seizures and neuropsychiatric symptoms [1,3,6,9,21,22,23]. A wide variation for the duration of these symptoms ranging from 1 day to 2 years has been documented in the literature [6]. However, although headache is widely known to be the most common among these symptoms and features as the presenting complaint in most of these patients [6,15,24], it can fluctuate and also be either progressive or permanent, hence making it not so sensitive and effective as the only single clinical parameter for monitoring these patients [1,3,8,25]. In addition, severity of the headaches does not correspond to the degree or extent of elevation in ICP[2]. In some cases, headaches may even be absent in the presence of severe intracranial hypertension [26,27]. The reason why some patients do not have headache even when their opening CSF pressures are often comparable to that of those who have headache is still unclear[14]. These challenges with headache are also coupled with the possibility of considerable overlap between the headache characteristic of raised ICP and the headache features of other primary headache disorders (such as migraine) or other disease conditions in patients with IIH [28,29,30,65]. Among the typical examination findings, papilloedema is one of the most common signs and has been well described [1,2,3,4,6,8,21,31,32]. It is also the most important sign [8]. Yet, the severity of papilloedema has not been found to be consistently related to visual loss [14,33,34,35]. Progressive visual loss in longstanding papilloedema and improvement of visual function in subsiding papilloedema can occur

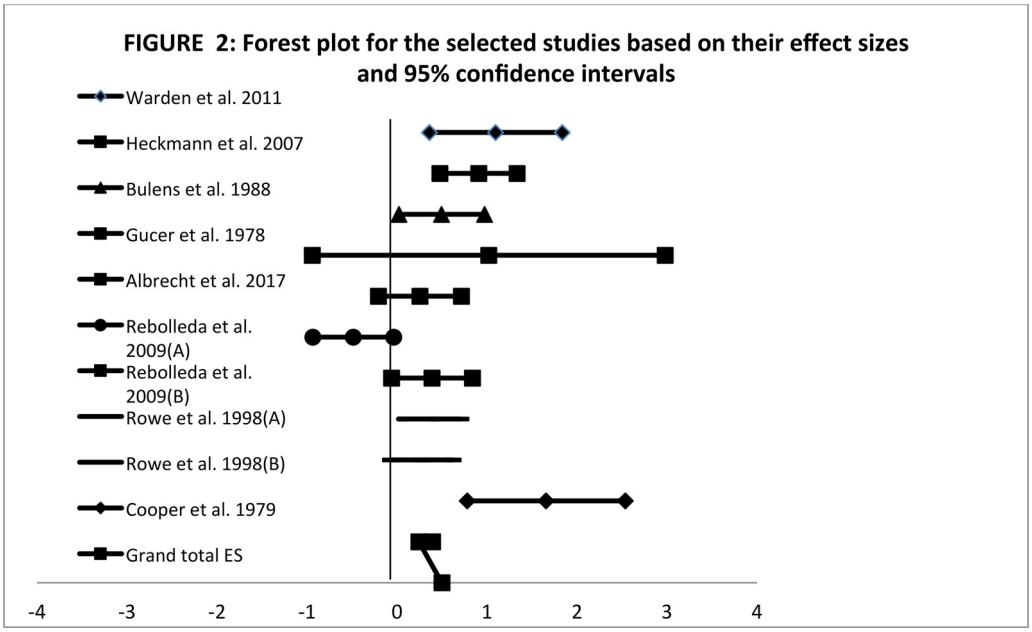

Fig. 2. Forrest plot for the selected studies based on their effect sizes and 95% confidence intervals.

without any change in visual acuity or visual field charting [34]. Furthermore, even though papilloedema is classical in IIH such that its absence in a suspected case should cause the diagnosis to be questioned, the disorder is well known to occur without any papilloedema in patients with headache and high CSF opening pressure [1,8,13,24,31,34,36,37]. This is commonly referred to as idiopathic intracranial hypertension without papilledema (IIHWOP) and had prevalence as high as 5.7% in one study [24,37,38]. There is no clear explanation yet for the occurrence of IIHWOP [24,35,36]. Meanwhile, there is no evidence-based work yet to demonstrate whether IIHWOP poses a threat to vision or not [13].

Furthermore, IIH has been known to be associated with cranial nerve palsies [1,2,4,8,25,39]. Abducens nerve palsy is generally the most common and occur in about 10% to 20% patients with IIH [7,13,25,32,39–41]. It could be either unilateral or bilateral [7,32,40]. It is also the most common neurological abnormality reported in 9–48% of children who have IIH [13,32]. In one series involving paediatric patients, the majority of cases who had abducens palsy also presented with papilloedema, higher BMI (body mass index) and opening pressure [32]. Luckily, it often resolves with appropriate and adequate treatment [32]. However, other cranial nerve palsies have been implicated though less frequently [25]. Olfactory, oculomotor, trochlear, trigeminal, facial, vestibulocochlear nerve and even hypoglossal palsies have all been reported [6,7,8,13,25,40,42–46]. Impaired function of some cranial nerves may also be involved in combination other cranial nerves due to IIH in the same patient [7,39,40]. Bilateral seventh nerve palsy has also been reported in connection with IIH [25,47,48] as well as bilateral vestibulocochlear nerve palsy [44] and bilateral oculomotor palsy [17]. Unfortunately however, all these could easily be mistaken as false localizing signs for other conditions and thus lead to potential wastage of valuable time and resources in directing investigation towards other such diseases which these patients could possibly have, while further damage from IIH continues [40,41,43]. Besides, some of them may be a nonspecific sign of elevated ICP from other causes [46]. In view of all these, the diagnostic principles section of the current guidelines recognizes no other cranial nerve involvement other than abducen palsy/palsies as a standard symptom of IIH [65].

4.2. Monitoring with ICP

Currently, there are conflicting views in the literature on the best method for this. Studies had previously demonstrated that intermittently raised ICP in IIH cannot be properly evaluated using just continuous ICP monitoring alone [13,31]. Continuous recording of CSF pressure has also been previously used to demonstrate that ICP may change over time in these patients and may reduce to normal levels even when the disease is progressive [21,22,49]. To compensate for these challenges, some have suggested repeating lumbar pressure measurements on periodic basis over time [13].

However, more recent studies have demonstrated that depending on lumbar puncture to ascertain intracranial pressure diagnosis and follow-up of patients with IIH may give spurious results and that direct

Table 3
Risk – of – bias assessment for selected studies.

Authors	Year of publication	Study population properly defined?	Outcomes and method of monitoring clearly defined?	Any loss of patients during follow-up?	Duration of monitoring clearly defined?	Regular interval for periodic assessment in course of monitoring mentioned?
Warden et al.	2011	Yes	Yes	No	Not clear	No
Heckmann et al.	2007	Yes	Yes	Yes	Yes	Yes
Bulens et al.	1988	Yes	Yes	No	Yes	No
Gucer et al.	1978	Yes	Yes	No	Yes	Yes
Albrecht et al.	2017	Yes	Yes	No	Yes	No
Rebolleda et al.	2009	Yes	Yes	No	Yes	Yes
Rowe et al.	1998	Yes	Yes	No	Yes	Yes
Cooper et al.	1979	Yes	Yes	Yes	Yes	Not clear

continuous ICP monitoring is indeed a more superior and accurate method of determining ICP than just depending on the opening CSF pressure [50–52]. Besides, regular measurement of CSF opening pressure is rarely used nowadays because the procedure is unpleasant [1]. Despite this advantage, direct continuous intraparenchymal ICP monitoring has only been suggested as a useful adjunct in the monitoring of IIH, and not as a sole method of monitoring [51,52]. Nevertheless, if clinical suspicion is sufficiently strong, repeat lumbar puncture is still justified if the initial opening CSF pressure is normal [13].

4.3. Monitoring with visual status

It has been recommended that current practice should focus on assessment of the symptoms, examination of the optic discs, measurement of visual acuity and formal perimetry to identify any visual loss, and then the visual fields documentated1 [15,53]. This is because visual impairment is the most serious challenge encountered with IIH [1,3,6,14,21,22,53]. However, this has been known to present in various patterns in terms of acuteness of its occurrence [1]. On one hand, it commonly causes gradual impairment of vision whereas in a smaller but significant number of patients, it has been known to follow a more rapid and aggressive course [1]. For those in whom it is gradual, the visual loss may go unnoticed until it becomes severe by the time it is detected [1]. Yet, another pattern is visual loss in patients in whom IIH recurs months to years after the original attack has resolved [1,14,24]. The attendant challenge regarding those in this third group is that although their recovery from the initial episode may be interpreted as resolved papilloedema, which is thought to be synonymous with the return of the pressure to normal levels, the CSF pressure can still be persistently increased for years even after that initial episode of clinical manifestation of the condition [32]. Meanwhile, papilloedema has been demonstrated to persist for years in IIH without serious visual impairment [34,54]. Furthermore, some patients with IIH develop visual failure without any headaches at all[14] and this is more common with children [14]. This varied pattern makes visual problems quite challenging as a sole method of monitoring IIH patients. A significant association was observed between visual loss and the occurrence of systemic hypertension in one study [1], while another study concluded that higher BMI may be directly linked to the increased risk of severe visual loss among patients with IIH [3]. These may be suggesting that co-morbid factors possibly play a major role in also potentiating this complication of the disease, in addition to effect from the disease condition itself on vision. Life style modification such as weight reduction, especially in overweight patients has been found to be beneficial for this set of patients based on evidence [8].

For these reasons, deterioration of vision alone has not been an ultimately acceptable monitoring tool for IIH [14]. However, it has been suggested that for asymptomatic patients with normal vision and just mild or no papilloedema, no treatment is needed and only serial ophthalmological evaluation is required [8]. For this group of patients, medical treatment has been advocated to be used first and surgical intervention only indicated if medical treatment fails or if their visual function is deteriorating [8].

4.4. Investigations for monitoring

Serial perimetry has been recommended as the most critical clinical test to obtain when following up these patients [15,55]. Thereafter, radiologic evaluation in form of fine high-resolution neuroimaging to show the optic nerve and disc may also be helpful [8,11,13]. Either thin section of CT scan sections or MRI imaging of the orbits may help to demonstrate both hydrops of the optic nerve sheath and also show if there is reversal of the optic nerve head [11,13]. Severe visual loss in patients with IIH has been well correlated with more frequent and more severe reversal of the optic nerve head using such neuroimaging[13]. Recent research with optical coherence CT has gone further to show that peripapillary retinal nerve fiber layer thickness and macular thickness coincides with and correlates with the severity of papilledema and may in fact be more sensitive than funduscopy not only in detecting optic nerve head elevation but also for monitoring the progression of the disease and for detecting its recurrence [11,12,56,57,58]. Additionally, 3-dimensional parameters of both the optic nerve head volume and the optic nerve head height measured using optical coherence CT has the capacity to differentiate between patients who have been treated and those who have had no treatment at all[56]. Other forms of neuroimaging such as Heidelberg retina tomography and Laser scanning tomography have been shown to be similarly sensitive and reproducible in the monitoring of optic disc swelling due to IIH [59,60]. Other studies have also suggested ultrasound (in B-mode) and high-resolution MRI scan to be a reliable, non-invasive tool to measure optic nerve sheath diameter in monitoring patients with IIH [61,62,63]. However, all these are merely findings from observational studies with no Class I evidence till date.

In view of all these pitfalls, it has been difficult to make any rigid recommendations on how monitoring of these patients should best be assessed [13].

5. Proposed risk stratification

5.1. Risk stratification on patient selection for monitoring disease progression in IIH

The section Q23 on monitoring IIH in the current guidelines [65] does not include ICP measurment as one of the parameters to be monitored [65]. The parameters also include funduscopy to grade papilloedema but however, exactly how the grading should be applied in monitoring is not specified or clearly defined [65]. Additionally, in the last recommendation under that section, outpatient review should be expedited if there is worsening of the visual fields or papilloedema but how worse both parameters should be for this to be done is also not specified [65]. On the basis of these observations coupled with above findings in the literature as highlighted, we recommend stratification of patients diagnosed with IIH to be considered for monitoring into the following four categories:

Category I - No additional risk / Low risk category: This should include -

- **patients with no demonstrable impairment of vision in either the short term (within 6 months) or long term (6 months or more) / Normal visual field status**
- **presentation with headaches associated with no abducens nerve palsy/palsies.**
- **patients with no papilloedema at all.**

Documentation of the optic nerve head with OCT (optical coherence tomography) to get a baseline for follow-up and thereafter, these patients should be monitored with funduscopy, visual field assessment but would not require ICP assessment with diagnostic LP. The frequency / interval of assessment should be every 3–6 months as outpatient, based on recommended follow-up intervals in the current guidelines [65]. The headaches should be characterized and managed as according to the guidelines [65].

Category II – Intermediate / Average risk category: should include patients with demonstrable but slow/gradual impairment of vision; patients with abducens palsy/ palsies.

- **In presence of demonstrable but slow or gradual impairment of vision / Visual field status affected but either improving or stable;**
- **Patients detected with either atrophic or mild papilloedema**
- **IIH with headaches associated with abducens palsy / palsies.**

For patients in this category, monitoring should also be with funduscopy, visual field assessment and should include diagnostic LP for initial assessment and follow-up but at a more frequent interval of 1–3 months, based on recommended follow-up intervals in the current guidelines [65]. Outpatient review should be expedited in this category of IIH patients if there is any objective demonstration of the visual field worsening or of papilloedema.

Category III -High risk category: should include patients with:

- **presence of rapid impairment of vision over a short period, in whom a putative higher risk of permanent visual loss may be anticipated / Visual fields significantly affected and worsening.**
- **Complete visual loss already at initial presentation**
- **In presentation with IIH associated with moderate or severe papilloedema**

Visual monitoring for this category should be as frequent as 1–4 weeks as per recommendation in the current guidelines [65] and should include ICP assessments by direct continuous intraparenchymal ICP monitoring rather than just LP for assessment of the opening CSF pressure. For those with total visual loss, either thin section of CT scan sections or MRI imaging of the orbits may help to demonstrate both hydrops of the optic nerve sheath and also show if there is reversal of the optic nerve head to help determine if the visual loss is still reversible. Monitoring within the hospital services may be advisable and required based on discretion of the managing physician.

Category IV – Indeterminate risk category: This should include

- **patients in whom IIH recurs months to years after the original symptoms have resolved** [24,64].
- **Patients with recurrence / persistence / progression of symptoms after surgical intervention**

Examples in this category should include patients with acute exercebation of headaches or recurrence of papilledema after medical therapy, CSF diversion or other surgical interventions (such as ONSF). Current guidelines recommends that papilledema be first ruled out in these patients using funduscopy65and if any demonstrable papilledema, should be investigated with lumbar puncture, CT scan or a shunt series [65]. If fundus is otherwise normal, they should be followed up with periodic ICP monitoring to rule out low-pressure headaches, since these headaches could possibly be as a result of CSF overdrainage following CSF diversion [65].

Irrespective of category for each patient, weight reduction, especially in overweight patients should be monitored periodically with the BMI. Treatment options and therapeutic intervention for each category should be as outlined in the current guidelines [65]. The ***presence of 2 or more parameters in each category*** (and not just a single parameter) should be the key determinant for the criteria each patient should belong to. Additionally, each patient may not necessarily progress stepladderwise from one category to the next.

6. Conclusion

We have suggested this proposed risk stratification strategy only as an addition to the current guidelines for follow-up and monitoring of IIH patients according to level of their risk of visual loss from the disease. We recognize the limitations of this study, particularly the findings in terms of pitfalls as highlighted and paucity of studies on the topic. Nevertheless, we are hopeful that this approach would be of potential clinical value and useful adjunct to the guidelines [65] for management of IIH.

Competing interests

The authors declare that they have no competing interests.

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Appendix A. Supplementary data

Supplementary material related to this article can be found, in the online version, at doi:https://doi.org/10.1016/j.clineuro.2019.02.013.

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