



Idiopathic Intracranial Hypertension Presenting with Sight Loss in a Young Woman: A Narrative Review of Medical, Weight-Management and Surgical Pathways

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Received: 16 January 2025; **Accepted:** 6 April 2025; **Published:** 5 August 2025

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Abstract

Idiopathic intracranial hypertension threatens sight in a population that is young, usually female, and usually well. The condition is treatable, the randomised evidence covers only mild visual loss, and the patients at greatest risk of blindness are the ones no trial has studied.

Objectives: *To review the published evidence on management of idiopathic intracranial hypertension: what the randomised medical evidence establishes, what is known about weight-directed treatment, when surgical intervention is indicated, and where the evidence does not reach.*

Material and Methods: *Narrative review of randomised trials, consensus diagnostic criteria, cohort studies, surgical series and society guidance indexed in PubMed and the major neurology and ophthalmology journals. No new patient data were generated; every estimate quoted is one published by the original investigators.*

Results: *The one randomised, double-masked, placebo-controlled trial enrolled 165 participants with mild visual loss, defined as a perimetric mean deviation between -2 and -7 dB, across 38 North American sites. All received a dietary intervention; those additionally allocated acetazolamide improved by 1.43 dB in perimetric mean deviation at six months against 0.71 dB with placebo, with improvements also in papilloedema grade, quality of life and opening pressure. The drug effect was independent of weight change. Sixty-nine of 86 in the acetazolamide group and 57 of 79 on placebo completed follow-up.*

Conclusion: *Acetazolamide has randomised support, and the effect is real but modest. Its evidence base excludes exactly the presentation that prompts urgent referral, since patients with severe or rapidly progressive visual loss were not eligible. For those patients the pathway is surgical and rests entirely on observational data.*

Abbreviations: *CSF – Cerebrospinal Fluid, IIH – Idiopathic Intracranial Hypertension, IIHTT – Idiopathic Intracranial Hypertension Treatment Trial, ONSF – Optic Nerve Sheath Fenestration, PMD – Perimetric Mean Deviation, VSS – Venous Sinus Stenting.*

Keywords: *Idiopathic Intracranial Hypertension; Papilloedema; Acetazolamide; Visual Loss; Cerebrospinal Fluid Diversion*

Introduction

Idiopathic intracranial hypertension is raised cerebrospinal fluid pressure without a mass lesion, hydrocephalus or an identified secondary cause. It presents with headache and papilloedema, and its importance lies in the papilloedema, because untreated it leads to progressive optic atrophy and permanent visual loss [1,2].

Diagnosis rests on the modified Dandy criteria as revised by Friedman, requiring symptoms and signs of raised pressure, normal neurological examination apart from cranial nerve palsies, normal neuroimaging including venography, elevated opening pressure on lumbar puncture and normal cerebrospinal fluid composition [3,4]. The population affected is predominantly female and predominantly of reproductive age, and incidence has risen in parallel with population weight [5,6,7].

Management has three strands: medical treatment to lower pressure, weight-directed treatment addressing the strongest modifiable association, and surgical intervention to protect vision when it is failing [8,9]. This review sets out the evidence for each and is explicit about which parts of it are randomised.

Material and Methods

Search strategy: PubMed and the archives of the major neurology, neuro-ophthalmology and neurosurgery journals were searched for studies of idiopathic intracranial hypertension. Search terms combined idiopathic intracranial hypertension and pseudotumor cerebri with acetazolamide, papilloedema, visual outcome, optic nerve sheath fenestration, shunt, venous sinus stenting and weight. Reference lists of consensus guidance [8,9] were screened by hand for primary sources.

Eligibility: Studies were included if they reported visual, headache or pressure outcomes in defined populations with the condition, established diagnostic criteria, or evaluated a medical, weight-directed or surgical intervention. Epidemiological studies are cited for context and identified as such.

Data extraction and handling: Design, sample size, entry criteria, severity of visual loss at baseline and the reported outcome were extracted as published. Baseline severity is reported alongside every treatment result, because the randomised evidence is confined to mild visual loss and applying it to severe presentations is the commonest error made with this literature. No pooling or re-analysis was performed. Figures 1 to 4 and Tables 1 and 2 reproduce published values only. This is a narrative synthesis and was not registered as a systematic review.

Results

The randomised medical evidence: One multicentre, randomised, double-masked, placebo-controlled trial has been conducted. It enrolled 165 participants with the condition and

mild visual loss at 38 academic and private sites in North America between 2010 and 2012, with six months of follow-up. All participants met modified Dandy criteria and had a perimetric mean deviation between -2 and -7 dB. All were offered a supervised dietary and lifestyle programme; participants were randomised to acetazolamide, escalated to the maximum tolerated dose, or matching placebo [10,11].

What it found: The primary outcome, change in perimetric mean deviation in the study eye from baseline to month six, improved by 1.43 dB with acetazolamide against 0.71 dB with placebo (Figure 1) [10,12]. Papilloedema grade, vision-related quality of life and lumbar opening pressure also improved significantly in the treatment arm, and the drug effect was reported as independent of weight change [10,12]. Sixty-nine of the 86 participants allocated acetazolamide completed follow-up (80 per cent) against 57 of 79 on placebo (72 per cent) (Figures 2 and 3) [11].

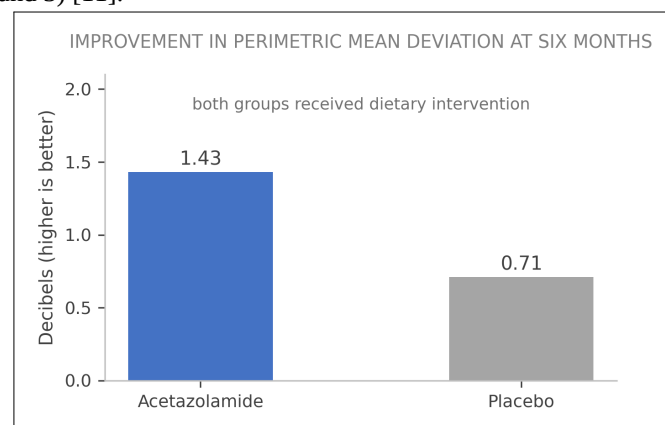


Figure 1: Improvement in perimetric mean deviation at six months, acetazolamide against placebo, both with dietary intervention [10,12]

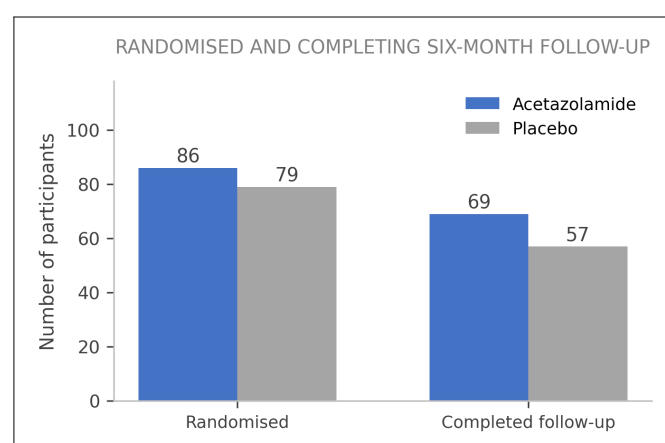


Figure 2: Participants randomised and completing six-month follow-up in the randomised trial [11]

How big is that effect: A difference of roughly 0.7 dB in perimetric mean deviation is statistically robust and clinically modest. It was obtained in patients whose visual loss was mild by design, on a scale running from 2 to -32 dB. The trial establishes that acetazolamide does something useful; it does not establish that it will halt a rapidly deteriorating field [10,13].

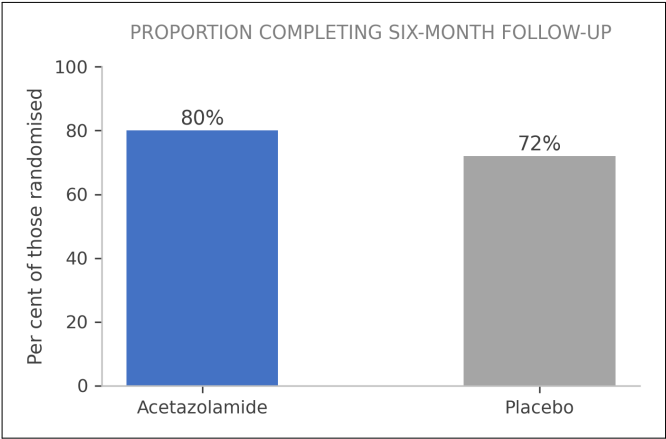


Figure 3: Proportion of randomised participants completing six-month follow-up [11]

What the trial excluded: Eligibility required a perimetric mean deviation between -2 and -7 dB (Figure 4) [11,14]. Patients with severe or fulminant visual loss, which is the presentation that generates emergency referral, were outside the trial and remain outside the randomised evidence entirely. Every recommendation for that group derives from observational series [15,16].

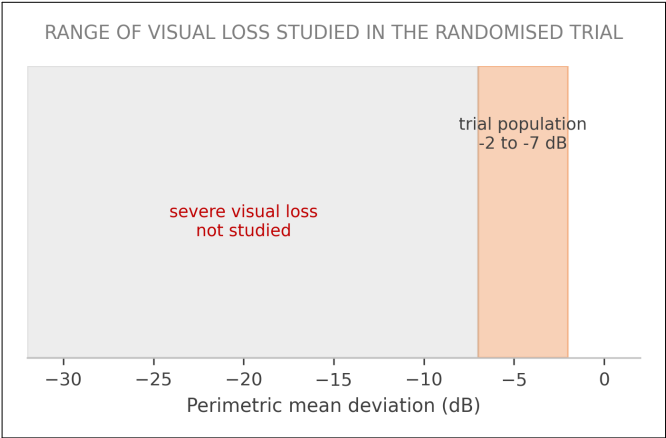


Figure 4: Range of visual field loss eligible for the randomised trial against the full scale of the measure [11,14]

Study	Design and cohort	Principal reported finding
IIHTT 2014 [10]	RCT, 165 participants, mild visual loss	PMD improved 1.43 dB vs 0.71 dB; papilloedema and QOL better
Wall et al. 2014 [11]	IIHTT design and methods	Entry restricted to PMD -2 to -7 dB; 38 sites
Friedman et al. 2013 [3]	Revised diagnostic criteria	Defines the diagnosis used throughout the literature
Mollan et al. 2018 [8]	Consensus guidelines	Sets out the management pathway including surgical thresholds
Mollan et al. 2021 [17]	Randomised trial of weight-directed treatment	Compared bariatric surgery with community weight management
Kalyvas et al. 2021 [18]	Systematic review of surgical options	Compares shunting, fenestration and venous sinus stenting
Thurtell 2019 [16]	Review of fulminant presentation	Management of rapidly progressive visual loss is observational

Table 1: Principal studies reviewed, with the finding as reported by the original investigators

Weight-directed treatment: Weight is the strongest modifiable factor associated with the condition, and weight reduction has been associated with reduction in intracranial pressure and improvement in papilloedema in cohort studies [19,20]. A randomised trial comparing bariatric surgery with a community weight management programme in women with active disease reported greater reduction in intracranial pressure in the surgical arm at 12 and 24 months [17]. This is an area where the clinical conversation is easily handled badly: the association is well established, patients frequently report that it is raised in a way that feels dismissive of their symptoms, and framing matters [21,22].

When vision is failing: Surgical intervention is indicated when visual function is deteriorating despite medical treatment, or when it is severe at presentation. Three approaches are used: cerebrospinal fluid diversion by ventriculoperitoneal or lumboperitoneal shunt, optic nerve sheath fenestration, and venous sinus stenting where a pressure gradient across a stenosis is demonstrated [18,23,24,25]. Comparative evidence between them is observational; a systematic review found all three associated with improvement in papilloedema and visual outcomes, with differing profiles of complication and revision [18,26]. Long-term review of shunting in this condition reports substantial revision burden [35]. Shunts have high revision rates, fenestration addresses vision more reliably than headache, and stenting is the newest and least studied [23,25,27]. No randomised comparison exists.

Headache: Headache is the symptom patients most often rank as dominant and is the one least reliably relieved by treatment directed at pressure. It frequently persists after papilloedema resolves and after pressure normalises, and often has migrainous features requiring separate management [28,29,30]. Treating the pressure and treating the headache are not the same task, and conflating them leads to repeated

procedures for a symptom that will not respond to them [31].

Intervention	Evidence status	Note
Acetazolamide	One randomised trial, mild visual loss only	PMD 1.43 vs 0.71 dB at six months [10]
Dietary and lifestyle programme	Given to both arms of that trial	Cannot be separated from the drug effect [10,11]
Bariatric surgery	Randomised against community programme	Greater reduction in intracranial pressure [17]
CSF diversion shunt	Observational only	Effective for vision; high revision rate [18,23]
Optic nerve sheath fenestration	Observational only	Vision protected more reliably than headache [24,26]
Venous sinus stenting	Observational only	Requires demonstrated pressure gradient [25,27]

Table 2: Interventions in idiopathic intracranial hypertension and the evidence supporting each

Discussion

The randomised evidence in this condition amounts to one trial. It was well designed, double-masked and placebo-controlled, and it demonstrated that acetazolamide improves visual field, papilloedema and quality of life over six months in patients with mild visual loss [10]. That is a genuine contribution to a field that had used the drug for four decades on the basis of physiological reasoning and small series [32]. A Cochrane review of interventions in this condition found the evidence base thin across the board [33].

Two things about it are worth stating precisely. The effect size is modest: 1.43 dB against 0.71, a difference of about 0.7 dB on a scale extending to -32 [10]. And both arms received a supervised dietary programme, so the trial answers what acetazolamide adds to that programme rather than what it does alone [11]. Neither point undermines the result; both change how it should be quoted.

The limitation that matters clinically is the entry criterion. Eligibility required mild visual loss, between -2 and -7 dB [11,14]. A young woman presenting with papilloedema and a field already worse than that, or deteriorating week by week, falls outside everything the trial established. For her, the evidence base is observational series and consensus opinion, and the decision is a surgical one taken under time pressure [15,16,18]. That inversion, where the evidence is strongest for the patients at least risk and weakest for those at greatest, is worth naming rather than glossing, and reviews of mechanism and management have made the same observation [34].

On weight, the evidence is clear and the communication is difficult. Weight reduction reduces intracranial pressure, and the randomised comparison of bariatric surgery against a community programme found the surgical route more effective on that endpoint [17,19]. Patients nonetheless report that the association is often raised in ways that feel like an explanation of why their symptoms do not warrant attention [21,22]. Both things can be true: the physiology is real, and the manner of its discussion determines whether the patient returns to clinic. In a condition

where the principal risk is irreversible sight loss from missed follow-up, that is not a soft consideration.

Study limitations

The limitations belong to the underlying literature. There is one randomised trial, of 165 participants, followed for six months, in North America, restricted to mild visual loss [10,11]. Long-term visual and headache outcomes beyond that window are observational. Both trial arms received a dietary intervention, so acetazolamide monotherapy against nothing has never been tested. Surgical comparisons are entirely observational, with selection determined by local expertise and by the severity that prompted intervention, which confounds any comparison between procedures [18,26]. Venous sinus stenting series are small and report short follow-up [25,27]. Diagnostic criteria have been revised during the period the literature spans, so cohorts are not strictly comparable [3,4]. Headache outcome measures are inconsistent across studies [28,30]. And almost all of this evidence comes from high-income settings [5,7].

Conclusion

For a young woman presenting with papilloedema and sight loss, the treatable component is the pressure and the thing to protect is the optic nerve. Acetazolamide has randomised support and produces a real if modest improvement, weight-directed treatment addresses the strongest modifiable association and now has randomised evidence of its own, and surgery protects vision when it is failing. The uncomfortable feature of this evidence base is its shape: the only randomised trial enrolled patients whose vision was mildly affected, while the patient who most urgently needs a decision is the one for whom nothing has been randomised. Recognising that should make clinicians quicker to escalate, not slower, because waiting for the medical treatment to work is a strategy the trial never tested in the patients most likely to lose their sight.

Acknowledgements

The authors thank the investigators whose trial and cohorts are drawn together in this article, and the participants who completed repeated perimetry over six months.

Funding

No dedicated funding from public, commercial or charitable sources supported the preparation of this review.

Conflict of Interest

The authors declare no competing interests.

Ethics Statement

This article reviews previously published studies only. No new patient data were collected and no participants were recruited, so ethical approval was not required.

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Citation: Doyle A, Bouchard N, Varga K. "Idiopathic Intracranial Hypertension Presenting with Sight Loss in a Young Woman: A Narrative Review of Medical, Weight-Management and Surgical Pathways." *J Clin Case Stud* (2025): 001-006 doi: 10.5281/zenodo.22237005