

## Benign intracranial hypertension diagnosed with bilateral papilloedema

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### Abstract

This article presents a case of benign intracranial hypertension (BIH) diagnosed from the presence of papilloedema. This potentially sight-threatening condition particularly affects younger obese females and can be idiopathic, caused by adverse reaction to certain prescription medications or by systemic disease. Prompt treatment is essential

to avoid optic atrophy and low energy diet and exercise forms part of long-term treatment to avoid relapse. Optometrists can play a critical primary health care role in the detection of papilloedema and referring appropriately.

**Key words:** Benign intracranial hypertension, papilloedema, obesity, headache

### Introduction

Benign intracranial hypertension (BIH) is often idiopathic<sup>1, 2</sup> or usually results in instances where cerebrospinal fluid reabsorption abnormality occurs, indicating venous system involvement<sup>1</sup>. Although relatively rare and a clinical entity of uncertain aetiology<sup>3</sup>, BIH manifests as elevated intracranial pressure in the absence of a brain tumour<sup>4</sup>, with an incidence<sup>5</sup> of approximately 19/100000. In most cases the diagnosis of increased intracranial pressure is confirmed by lumbar puncture<sup>6</sup>. Whilst, in the absence of a brain tumour, the condition may be described as benign, it is certainly dangerous and poses a significant threat to the visual prognosis<sup>4</sup>. BIH is also known as *pseudotumour cerebri*<sup>2, 7</sup>; a name deriving from raised ICP and papilloedema in the absence of a tumour. Many clinicians however prefer

to use the term idiopathic intracranial hypertension and consider the disorder as a diagnosis of exclusion<sup>2</sup>. To regard the condition as benign is rather misleading as relapse after treatment is common and untreated sequelae are potentially visually disastrous<sup>8</sup>. The condition may be self-limiting or become chronic with substantial vision loss and headache resulting in significant morbidity<sup>5</sup>.

The majority of BIH patients appear to be young<sup>8</sup> or of child bearing age, female<sup>7, 8, 9</sup> and obese<sup>8, 10, 11</sup> with headaches a common generally reported symptom<sup>3, 12</sup>. Black patients seem more likely to have more risk factors like hypertension, sleep apnoea, obesity and anaemia with consequently more severe visual loss in at least one eye than patients<sup>13</sup> with lightly-pigmented skin. Characteristic ocular symptomatic features of BIH include reduced visual acuity, diplopia and some visual field restriction<sup>3</sup>. Clinical signs include

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optic disc swelling (papilloedema) usually, but not exclusively, bilaterally<sup>9</sup> with or without haemorrhaging depending on the severity of the condition and diplopia from possible abducens nerve involvement<sup>14</sup>. Ocular symptoms have been shown to exist in more than 90% of cases of BIH<sup>9</sup> and the condition is an important source of headaches in the paediatric population, including the non-obese, potentially leading to irreversible blindness<sup>12</sup>.

### Case History

This case study stems from GF, an overweight 35 year-old coloured female home-carer, who presented for a consultation on Friday, 15 October 2010 at 14.30. Her chief complaint was of deteriorating vision and non-specific headaches. Further detailed questioning regarding her headaches elicited a frequency of mild yet constant dull ache of a few weeks duration. The precise location of the headache was unclear with no obvious associated activity and no need for analgesic medication. She reported no significant medical history and had never before had an eye examination and was on no chronic medication.

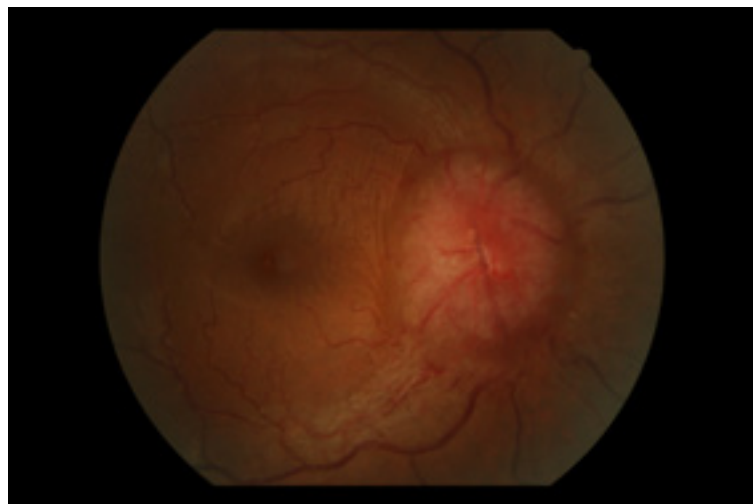
### Clinical assessment and findings

Her unaided visual acuity measured with the Snellen chart at 6 metres was 6/7.5<sup>-2</sup> in the right and 6/9<sup>-2</sup> in the left eye. Confrontation fields appeared slightly contracted and slit lamp examination was unremarkable, with adequate tear break-up time. Pupil assessment and ocular motilities including the cover test exhibited no abnormalities. Fundoscopy revealed narrowed blood vessels with prominent arterio-venous crossings and swollen optic discs, left more than right, accompanied by bilateral haemorrhaging again left more than right, which was further confirmed by fundus photography (Figures 1 and 2).

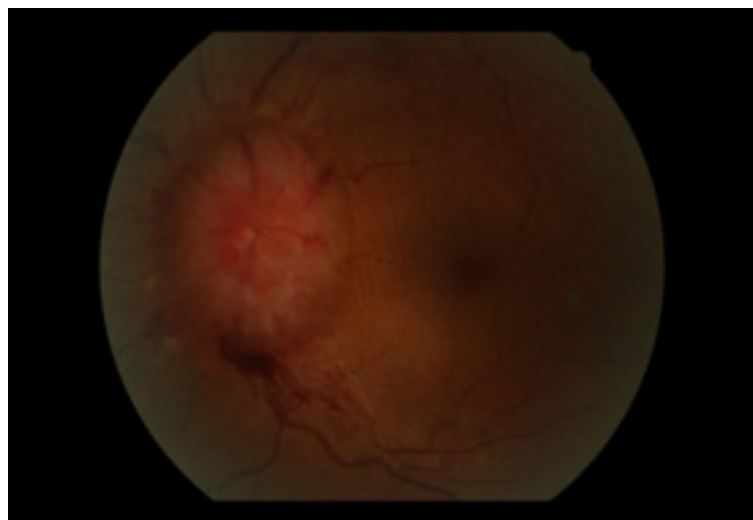
### Management

GF was referred to an ophthalmologist and was seen on the following Monday, where the diagnosis of bilateral optic disc oedema was confirmed. Her blood pressure was measured at 164/100 mmHg and the presence of bilateral Bjerrum scotomata was also confirmed. She was referred to a neurologist for

admission to hospital and an MRI scan. The MRI scan proved to be normal but a lumbar puncture confirmed benign raised intracranial pressure. The patient was placed on acetazolamide (Diamox) and the bilateral papilloedema subsequently resolved. By the time of her ophthalmological follow-up three months later, her vision had reverted to 6/6<sup>-1</sup> OU with full fields.



**Figure 1.** Patient GF: Right swollen disc and smaller inferior haemorrhage (Fundus photograph by KC Phillips)



**Figure 2** Patient GF: Left swollen disc with larger inferior haemorrhage (Fundus photograph by KC Phillips)

### Discussion

Papilloedema describes a swelling of the optic disc resulting from intracranial pressure transmitted to the subarachnoid space adjacent and surrounding the optic nerve<sup>3</sup>. The absence of a venous pulse is often cited as pathognomic of papilloedema<sup>15</sup>. Paradoxically, some patients have been reported

as having papilloedema with normal intracranial pressure leading some researchers to suggest that papilloedema does not always occur solely as a result of raised intracranial pressure but sometimes by a sealing of the subarachnoid space of the optic nerve causing a toxic environment around the nerve<sup>3</sup>. Notwithstanding the pathophysiology of the condition, the clinical manifestation of papilloedema with BIH as the underlying cause can be found in certain pathologies including the following:

#### *Prescription drug use*

An association has been observed between idiopathic intracranial hypertension and minocycline use<sup>16</sup>, where the initial treatment is for acne, often occurring weeks after commencement of the medication. Vitamin A and isotretinoin (Accutane) as acne treatment have also been linked, along with acitretin, and etretinate, to intracranial hypertension with significant ocular side effects<sup>17</sup>. Idiopathic intracranial hypertension has been clearly associated with both steroid withdrawal and anabolic steroid use<sup>14, 18</sup>, thyroid replacement therapy in children and the consumption of lithium<sup>14</sup>. Tetracycline is a further probable cause of intracranial hypertension<sup>14</sup> which is problematic due to its use in the treatment of infectious diseases causing the condition.

#### *Malignant hypertension*

Malignant hypertension (MHT) is a clinical and pathological syndrome defined<sup>19</sup> by a marked increase in blood pressure in the region 180/120 mmHg. MHT is accompanied by retinal haemorrhage and exudates, with or without papilloedema and it can present as an acute renal or cardiac failure with hypertensive encephalopathy. In such cases, severe hypertension demonstrates its immediate damaging potential due to malignant vascular injury and disruption of vascular endothelium<sup>19</sup>. Malignant hypertension may develop in patients with essential or secondary hypertension and should be excluded when evaluating BIH.

#### *Systemic Lupus Erythematosus (SLE)*

Idiopathic intracranial hypertension (IIH) has been found to account for a considerable number of cases of intractable headaches in SLE patients, which is relieved by the use of steroid medication<sup>20</sup>. The likely cause is venous sinus thrombosis<sup>14</sup>.

#### *Syphilis*

Syphilis is a rare but clinically important differential diagnosis in cases of IIH, especially in nonobese patients, where a syphilitic central nervous system infection can give rise to the condition. Syphilis is usually resolved with appropriate antibiotic treatment<sup>21</sup>.

#### **Prognosis**

Grading of papilloedema has become easier with the widespread use of the Optical Coherence Tomography (OCT)<sup>15</sup>. Where previously the severity of the condition was graded as 0 (absent), grade 1 (mild), grade 2 (moderate) and marked (grade 3) utilising the Frisén grading scale for papilloedema in conjunction with stereoscopic photographs<sup>22</sup>, now the degree of optic disc swelling can be accurately quantified with the OCT<sup>15</sup>.

Relapse after treatment occurs in approximately 28% of patients<sup>8</sup>. Relapse incidence has been reduced by the implementation of long-term therapy in the form of low-energy diet and moderate exercise. Loss of weight appears to result in complete resolution of both the BIH and papilloedema, even three months after the diet has been stopped<sup>23</sup> where weight loss had been maintained. Whereas headache was determined to be a poor indicator of active disease, weight gain was often found in those suffering relapse, compared to those in the non-relapse group<sup>8</sup>, indicating that obesity is a compelling risk factor for BIH. Treatment of other co-morbidities like hypertension and sleep apnoea, which have also been shown to be risk factors<sup>13</sup>, should further reduce the danger of relapse.

The immediate treatment of BIH is usually with acetazolamide<sup>9,24</sup> while weight loss has been associated with a complete resolution of low-grade papilloedema and should be utilised as a long-term therapy<sup>25</sup>. Low-energy diet has been associated with marked reduction in intracranial pressure, improvement on headache impact test score, papilloedema (as measured by ultrasonography and OCT), mean improvement in visual field and visual acuity<sup>23</sup>.

Whilst BIH is a relatively rare occurrence, the threat to vision is a very real one. In the light of the apparent worldwide increase in obesity, a known risk factor for BIH<sup>7-9</sup>, and the apparently more serious visual outcomes for Black patients<sup>13</sup> with BIH it would be prudent for all optometrists, particularly in South Africa, to be especially vigilant for signs of

the condition. Any complaint of headaches should be fully investigated as a matter of course and those of a non-specific nature should not be dismissed out of hand. The everyday use of the direct ophthalmoscope as well as other indirect retinal-viewing instruments, including fundus cameras, places the optometrist in a critical primary healthcare position in the detection of papilloedema resulting in the diagnosis of intracranial hypertension with certainly vision-saving and possible life-saving consequences. With increasing use of OCT and ultrasonography by optometrists this valuable primary-care service can be expanded and facilitate more appropriate referral to the relevant medical specialist.

## Conclusion

The risk factors for BIH include obesity and female gender of reproductive age which would include a significant number of people in South Africa. Hypertension and sleep apnoea are further risk factors and often co-morbidities such as SLE and syphilis are present. Black patients appear to suffer poorer visual outcomes and the use of certain prescription medications including those commonly used in the treatment of infective and inflammatory conditions as well as acne can give rise to the condition. Optometrists should bear in mind the risk factors and assess each patient on an individual basis evaluating potential areas of concern whilst eliciting a thorough case history. Prompt referral to ophthalmology or neurology is essential in order to facilitate the resolution of the condition.

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