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*Research Article*

# **Idiopathic Intracranial Hypertension in a Mother and her Son, is it Familial or Environmental?**

**Authors**

**Khalid Mohamed<sup>1</sup>, Amira Saeed<sup>2</sup>, Abdulla Ehlayel<sup>2</sup>, Khalid Ibrahim<sup>1</sup>,  
Mahmoud F Elseid<sup>1</sup> and Yahia Z. Imam<sup>3</sup>**

Departments of Paediatric Neurology<sup>1</sup>, Paediatrics<sup>2</sup> and Neurology<sup>3</sup>  
Hamd Medical Corporation, Doha, Qatar

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## Abstract

**Introduction:** Idiopathic intracranial hypertension (IIH) is defined as intracranial hypertension in the absence of other intracranial pathology such as space occupying lesion or CNS infection based on clinical, radiological and laboratory evaluation. **Objective:** To report on a mother and son with IIH who also share the environmental factors of obesity and family distress. **Results :** A 32 year old woman presented with headache and visual symptoms, her 9 year old son started complaining of similar symptoms around three months later .They were both obese with a body mass index (BMI) of over 35, the mother was also treated for depression and there were recent family distresses. Examination revealed papilloedema; CSF pressure was high on LP. They were both treated successfully with

Acetazolamide with resolution of the symptoms and improvement of the papilloedema. **Discussion:** Presentation with Idiopathic intracranial hypertension in the same family is rare; in many cases clear environmental factors such as corticosteroid treatment or the use of antidepressants were identified as possible causes. **Conclusion:** We report a rare presentation of IIH in a family with a mother and son who were treated successfully with Acetazolamide.

**Keywords:** Idiopathic intracranial hypertension, familial, obesity

## **Case reports**

### ***Case1:***

A 32 year old lady who suffers from depression for the last year was on treatment with Paroxetine she also reported a history of oral contraceptive use. She presented with a three months history of headache, which she describes as bilateral frontal with moderate intensity lasting on average for around two hours every two to three days ; the headache is aggravated by bending forward and is occasionally associated with visual symptoms mainly blurring of vision and improves partially upon resting and use of analgesia.

On examination: she was well looking with normal vitals signs and was obese with a body mass index (BMI) of 39.

The neurological exam was normal apart from bilateral papilledema; formal perimetry was normal.

Routine hematology and biochemistry investigations and thyroid function tests were normal.

CT with contrast showed only empty sella turcica and no evidence of venous thrombosis or space occupying lesions. This was confirmed on MRI which included MRV to exclude venous thrombosis, both were reported as normal.

Lumbar puncture showed a high opening pressure of 55 cm H<sub>2</sub>O.



Cerebrospinal fluid (CSF) analysis showed normal cell count, normal glucose, protein and cytology.

CSF was negative for routine gram stain and cultures as well as tuberculosis (TB) PCR and TB culture.

The patient was labeled as idiopathic intracranial hypertension and was started on acetazolamide 500 mg twice daily. Paroxetine was tapered and stopped following discussion with the treating psychiatrist. She was given life style advice to lose weight.

The patient was seen 4 months later; she reports resolution of her headaches. Fundoscopy showed remarkable resolution of her papilledema, the rest of the neurological examination was normal.

## ***Case 2:***

A nine year old boy, the eldest son of the patient mentioned above, presented following his mother's presentation with headaches; the headaches started around four months prior to presentation, shortly after the onset of his mother's headache. His headaches were severe and dull and lasted for a few hours without nausea or vomiting. They used to occur around twice to three times per week. There was no history of late night or early morning headaches and no other neurological symptoms. His mother also noted that his school work was not as good as the year before.

On examination, he was found to be well with normal blood pressure; he was overweight with BMI of 32, his neurological examination was normal apart from bilateral papilloedema which

was confirmed by formal ophthalmology examination. His visual fields and acuity were normal although he does wear glasses for short-sightedness.

He underwent an MRI scan which was reported as normal including MRA and MRV; he underwent LP which revealed CSF pressure of 38 CM of water, this was reduced to 18 by CSF withdrawal. CSF analysis showed normal protein, glucose and lactate; seven white cells and negative bacteriology, virology screening and TB PCR.

He was commenced on Acetazolamide 250 mg twice daily; six weeks later, at follow up, he showed marked improvement in the papilloedema and resolution of the headaches.

## **Discussion**

Idiopathic intracranial hypertension (IIH) is defined as intracranial hypertension in the absence of other intracranial pathology such as space occupying lesion or CNS infection based on clinical, radiological and laboratory evaluation.

IIH is preferred to as Benign Intracranial Hypertension as the condition is not benign and can lead to significant morbidity and visual loss.

There have been several reports of familial IIH including a case report from the UK by Beri et al describing a mother and 2 daughters with variable phenotypes.

A detailed report by Corbett (2008) in the journal of neuro-ophthalmology which reviewed a large cohort of patients and described 11 families with more than one affected individual most affected members were mother and daughter and 8 out of his 11 families had a significant history of severe obesity.

The third report we identified was by Traviesa et al from 1976 describing three sisters with IIH; they were all obese.

Our case report highlights that common environmental factors such as obesity as well as genetic factors may play an important role in the etiology of IIH which is a common serious neurological condition presenting with headaches and visual symptoms. This is as far as we are aware the first case report of familial IIH in the

Middle East and the first to report the condition in a mother and son.

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