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Mitchell, James; Mollan, Susan; Tsermoulas, Georgios; Sinclair, Alex

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Telemetric monitoring in Idiopathic Intracranial Hypertension demonstrates intracranial pressure in a case with sight threatening disease

Authors: James L Mitchell MRCP^{1,4}, Susan P Mollan FRCOphth², Georgios Tsermoulas FRCS³, Alex J Sinclair PhD^{1,4,5}

- 1 Metabolic Neurology, Institute of Metabolism and Systems Research, University of Birmingham, Birmingham, United Kingdom (UK)
- 2 Birmingham Neuro-Ophthalmology, Queen Elizabeth Hospital, Birmingham, UK
- 3 Department of neurosurgery, University Hospitals Birmingham, Queen Elizabeth Hospital, Birmingham, UK
- 4 Department of Neurology, University Hospitals Birmingham, Queen Elizabeth Hospital, Birmingham, UK
- 5 Centre for Endocrinology, Diabetes and Metabolism, Birmingham Health Partners, Birmingham, UK

Corresponding Author: Miss Susan P Mollan, Birmingham Neuro-Ophthalmology, Queen Elizabeth Hospital, Birmingham, UK. Susan.mollan@uhb.nhs.uk 0000-0002-6314-4437

Abstract

The understanding of raised intracranial pressure (ICP) is increasing with the directed use of intracranial telemetric ICP monitors. This unique case observed ICP changes in a patient with idiopathic intracranial hypertension (IIH), who developed rapid sight-threatening disease. A lumbar drain was inserted, as a temporising measure, and was clamped prior to surgery. This resulted in rapid rise in ICP, which normalised after insertion of a ventriculoperitoneal shunt. This case highlighted the utility of the ICP monitor and the lumbar drain as a temporising measure to control ICP prior to a definitive procedure as recommended by the IIH consensus guidelines.

Key Words

Pseudotumor cerebri; papilledema; raised intracranial pressure; headache; lumbar drain; telemetric monitor

Abbreviations

CSF	Cerebrospinal fluid
LP	Lumbar puncture
IIH	Idiopathic intracranial hypertension
ICP	Intracranial pressure

Introduction

Idiopathic Intracranial Hypertension (IIH) is characterized by increased intracranial pressure (ICP) with no identifiable cause. [6] Also known as pseudotumor cerebri, it occurs mainly in overweight women of working age. [1,6,10,] There is a rising incidence and prevalence in this disease. [1,10] Headache is a major morbidity [16] and initially this is progressively more severe and frequent, with a divergence of traditional considerations of a raised intracranial pressure headache [15] to a phenotype that is highly variable and commonly mimics migraine. [12] For those with declining visual function the typical treatment is emergency surgery with either cerebrospinal fluid (CSF) diversion or optic nerve sheath fenestration. [7,11]

There is much to understand regarding CSF dynamics in IIH, as long-term data is emerging. [18,19] Lumbar puncture (LP) remains the commonest way to indirectly measure intracranial pressure. However LPs cause local discomfort, low pressure headaches and more rarely infection or local haemorrhage. [5,20] It is recognised that LP induces a transient reduction of CSF pressure, although the effect is typically short lived with pressures found to rise rapidly after the procedure, despite the amount of CSF drained. Those with IIH who have undergone LP frequently recall negative and emotional experiences subsequently. [17]

External ventricular drains and intraparenchymal monitors are typically used to monitor intracranial pressure, however long-term use is limited due to the risk of infection. Telemetric ICP monitoring has increasing clinical utility to enhance decision making in complex cerebrospinal fluid (CSF) disorder cases. [2,4] Within the IIH community of professionals and patients, who undertook a priority setting partnership, the aim was to understand the disorder more comprehensively and to recognise when surgical intervention is warranted. [9]

Case presentation

A 39 year old Caucasian woman presented to the neuro-ophthalmology service with papilledema. Eight years earlier she had been diagnosed with IIH which had remitted and she was asymptomatic for the prior six years. On presentation her symptoms had returned with pulsatile tinnitus; daily headache of a migraine-like phenotype with visual aura; visual blurring and transient visual obscurations on coughing or straining. On examination her visual acuities were 6/6 in both eyes, colour vision was full and she had no afferent pupillary defect. On dilated examination she had moderate papilledema (frisen grade 2 bilaterally) (Fig. 1a,b). Her body mass index at presentation was 33kg/m², she reported significant weight loss following the initial diagnosis (although not documented clinically) in conjunction with disease remission. Upon recurrence of her IIH her neuro-imaging including CT head and CT venography, excluded venous sinus thrombosis, space occupying lesion and hydrocephalus. Lumbar puncture documented the opening pressure was at 35cm CSF, with normal constituents.

She was recruited to a research study where a telemetric intracranial pressure (ICP) monitor (Raumedic™ p-Tel) was inserted (Fig. 2a) and the ICP was recorded over prolonged periods during home monitoring (Fig. 3a). Over the 10 days post-surgery (prior to initiation of any trial specific intervention) she noticed an increasing number of daily transient visual obscurations and sought reassessment. Examination noted worsening of her papilledema (Fig. 1c,d) and visual field deterioration, including bilateral enlarged blind spots and a left central positive scotoma (Fig. 4 c,d). There was no weight increase over this short period between this presentation and worsening of her symptoms. The disease had progressed to fulminant IIH, at this point the ICP trace shows a markedly raised ICP and the presence of a-waves, suggesting a critical reduction in intracranial compliance (Fig. 3b). During these a-waves, she reported excruciating holocranial head pain, on a verbal rating scale of 10/10 (where 10 is the most severe). The histogram (Fig. 5) shows the shift in ICP between baseline and the fulminant phase. A lumbar drain was inserted as a temporising measure, in accordance with published guidelines [7,11], whilst awaiting a definitive neurosurgical intervention. The trace, during the lumbar drain insertion, shows a marked drop of ICP at the point of insertion (Fig. 3 c). The lumbar drain was clamped, 3 days after insertion, before the procedure to facilitate insertion of the ventricular catheter in the slit ventricles. The lumbar drain had not reversed the underlying disease process and ICP rose from mean -2.0mmHg (equivalent to 2.7cmCSF) to mean 40.6mmHg (equivalent to 55.2cmCSF) over 2 hours (Fig. 3 d). The ICP post-operatively settled, mean 18.9mmHg (equivalent to 25.7cmCSF) (Fig. 3 e).

Discussion

The case reports direct measurement of ICP in a patient with IIH who deteriorated, requiring surgical intervention to preserve vision. The ICP recordings demonstrate several new findings which have not, to the authors knowledge, been reported previously. First, a peak of ICP during an a-wave of 76mmHg, with a mean ICP of 60mmHg over a 10-minute period (Fig. 3 b). During these peaks the patient reported very severe headache on a verbal rating scale of 10. As the shunt insertion could not happen within 24 hours a lumbar drain was inserted, as recommended by IIH guidelines.[7,11] During this recording we see a mean ICP of 14.7mmHg (Fig. 3 c), demonstrating the effectiveness of the drain in controlling ICP. Secondly, when the drain is clapped, just prior to surgery to effect filling of the ventricles, the ICP rapidly rises within 2 hours to a mean of 40.6mmHg (Fig. 3 d). This last point demonstrates that the disease was not put into remission by prolonged drainage in this case.

Debate exists in the IIH literature regarding lumbar puncture as a method of treatment.[14] Little is known about the spectrum of disease in IIH, and certainly this case, which is likely to represent the more severe end, given the ICP recordings, did not remit despite prolonged lumbar drain. At the outset of disease or relapse there are currently no clinical markers to allow the clinician to decide which patients are at risk of imminent visual loss, therefore the consensus guidance does not recommend lumbar puncture as a treatment.[7,11]

The most common method of ICP measurement is lumbar puncture, but this is limited as it measures a one-off measurement with side effects including discomfort of the procedure. [5,17,20] Direct measurement of ICP can either be non-invasive or invasive. A reliable and validated method of non-invasive ICP measurement is yet to be deployed in routine clinical use.[8] Invasive measurement of ICP are often not warranted in this disease, as there is little evidence to suggest absolute pressure measures drive clinical decision making. The use of wireless or telemetric ICP monitors can be helpful. There are 2 main systems available commercially at present, Neurovent p-Tel™ (Raumedic, Helmbrechts, Germany) and Sensor Reservoir (Miethke™, Potsdam, Germany). The Neurovent™ p-Tel sites a pressure sensor in the brain parenchyma, in contrast the Miethke™ system places a sensor within a reservoir (chamber containing drained CSF mounted external to the skull under the scalp) attached to a ventricular drain thus reading intra-ventricular pressure. The complication rate for the Raumedic p-Tel is 6% overall, with seizures affecting 3% and infection in 1.5%.[3] It should however be noted that this was in a series of patients with significant structural brain abnormality and pathology (hydrocephalus, trauma and haemorrhage) and the rate of such complications is likely lower in IIH patients. The Raumedic™ p-Tel device provides a high degree of accuracy, many devices have been kept in situ beyond the licensed 3-month period where they have been shown to have drift of 2.5mmHg (3.2cm H₂O) over median 8 months and in some patients used for up to 5 years.

Telemetric ICP monitors have an evolving role in diagnosis and monitoring of CSF disorders. In complex cases ICP monitoring might facilitate the decision to proceed to neurosurgical shunt placement, in a patient developing fulminant IIH. The ICP monitors can improve diagnostic accuracy in atypical or complex cases such as those with Chiari (with contra-indications for LP) and in those whose disease might be complicated by concurrent CSF leak. The ICP monitors have utility in shunted patients to refine shunt settings, exclude shunt malfunction and avoid over-drainage symptoms, which are noted in up to a quarter of patients. ICP telemetry may also facilitate the differentiation between raised pressure headaches and non-pressure related headaches such as migrainous headaches or medication over-use headache. This is particularly important in IIH, where exacerbation of headache in the shunted patient can be complicated to investigate.[13]

Conclusion

Telemetric ICP monitors are increasingly being used to manage difficult cases of IIH (although in this patient the monitor was originally sited for research). Deciding if and when to escalate to surgical intervention can be a challenge in IIH. There are no trials to guide our practice currently. In this case the ICP monitor corroborated the clinical findings and empowered the correct, timely, decision to move to surgical intervention. In the future, greater knowledge of the relationship between high spiking ICP pressures and optic nerve damage may provide information on the timing of surgical intervention in IIH. Lumbar drainage is a useful holding measure whilst awaiting a definitive procedure to lower ICP, but did not reverse the disease process in this patient with fulminant disease.

Compliance with ethical standards

Informed consent

The patient consented to the submission of the case report to the journal.

Conflicts of Interest

JLM- Invex therapeutics, consulting (2020)

SPM -Invex therapeutics, advisory board (2020)

GT- None

ASJ - Invex therapeutics, company director with salary and stock options (2019, 2020)

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All authors have read and approved the final manuscript.

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Fig. Legend

Fig. 1 Optical coherence tomography (OCT) imaging (Heidelberg Engineering, Heidelberg, Germany) infrared reflectance fundal images demonstrating changes in papilledema over the clinical course.

- a) right eye b) left eye at baseline;
- c) right d) left at fulminant presentation;
- e) right, f) left 6 weeks post VP shunt with complete resolution of the papilledema

Fig. 2

- a) X-radiograph of skull showing telemetric Rau monitor
- b) X-radiograph of skull showing telemetric catheter and ventricular shunt placement

Fig. 3

Telemetric intracranial pressure monitoring in a case of fulminant IIIH

- a) Baseline ICP trace, mean ICP 23.7mmHg range 14.0-58.0mmHg.
- b) ICP trace during admission for fulminant IIIH prior to intervention, mean ICP 33.3mmHg, range 9.2-76.0mmHg, period marked 'A' wave corresponded to a mean 76mmHg, highest maximal peak recorded during this period was 111.3mmHg (not shown).
- c) ICP trace during lumbar drain insertion (arrow), mean pre-41.5mmHg range 15.4-70.6mmHg, post insertion mean 14.7mmHg, range 1.8-40.0mmHg.
- d) 2-hour ICP trace recorded after closure of lumbar drain, initial mean -2.0mmHg, shaded area mean 40.6mmHg range 26.6-77.0mmHg.
- e) ICP trace recorded post-op ventricular-peritoneal shunt insertion, mean 18.9mmHg, range 8.6-36.6mmHg.

All ICP monitoring shown was in the supine position, ICP trend data shown at 1Hz, 1hr trace shown aside panel D) 2 hrs. 1mmHg converts to 1.36cm H₂O. Mean and range automatically calculated by software, Dataview version, Raumedic, Helmbrechts, Germany

Fig. 4

Grey scale of the Humphrey visual fields.

- a) right eye at baseline showing enlarged blind spot and nasal step;
- b) left eye at baseline showing enlarged blind spot;
- c) right eye at fulminant presentation increase in the nasal step;
- d) left eye at fulminant presentation increase in the blind spot;
- e) right eye 6 weeks post VP shunt with complete resolution of the papilledema shows an inferior defect in the visual field;
- f) left eye 6 weeks post VP shunt with complete resolution of the papilledema however the enlarged blind spot remains

Fig. 5

Histogram showing ICP frequency at baseline and the shift in ICP observed with the fulminant presentation