Telemetric monitoring in idiopathic intracranial hypertension demonstrates intracranial pressure in a case with sight-threatening disease

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Abstract

The understanding of raised intracranial pressure (ICP) is increasing with the directed use of intracranial telemetric ICP monitors. This case uniquely observed ICP changes by telemetric monitoring 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 a 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.

Keywords 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. Also known as pseudotumor cerebri, it occurs mainly in over-weight women of working age [1, 10]. There are 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 to a phenotype that is highly variable and commonly mimics migraine [13]. For those with declining visual function, the typical treatment is emergency surgery with either cerebrospinal fluid (CSF) diversion or optic nerve sheath fenestration [6].

There is much to understand regarding CSF dynamics in IIH, as long-term data is emerging [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 [18].

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 [3, 4]. Within the IIH community of professionals and patients, who undertook a priority setting partnership, the

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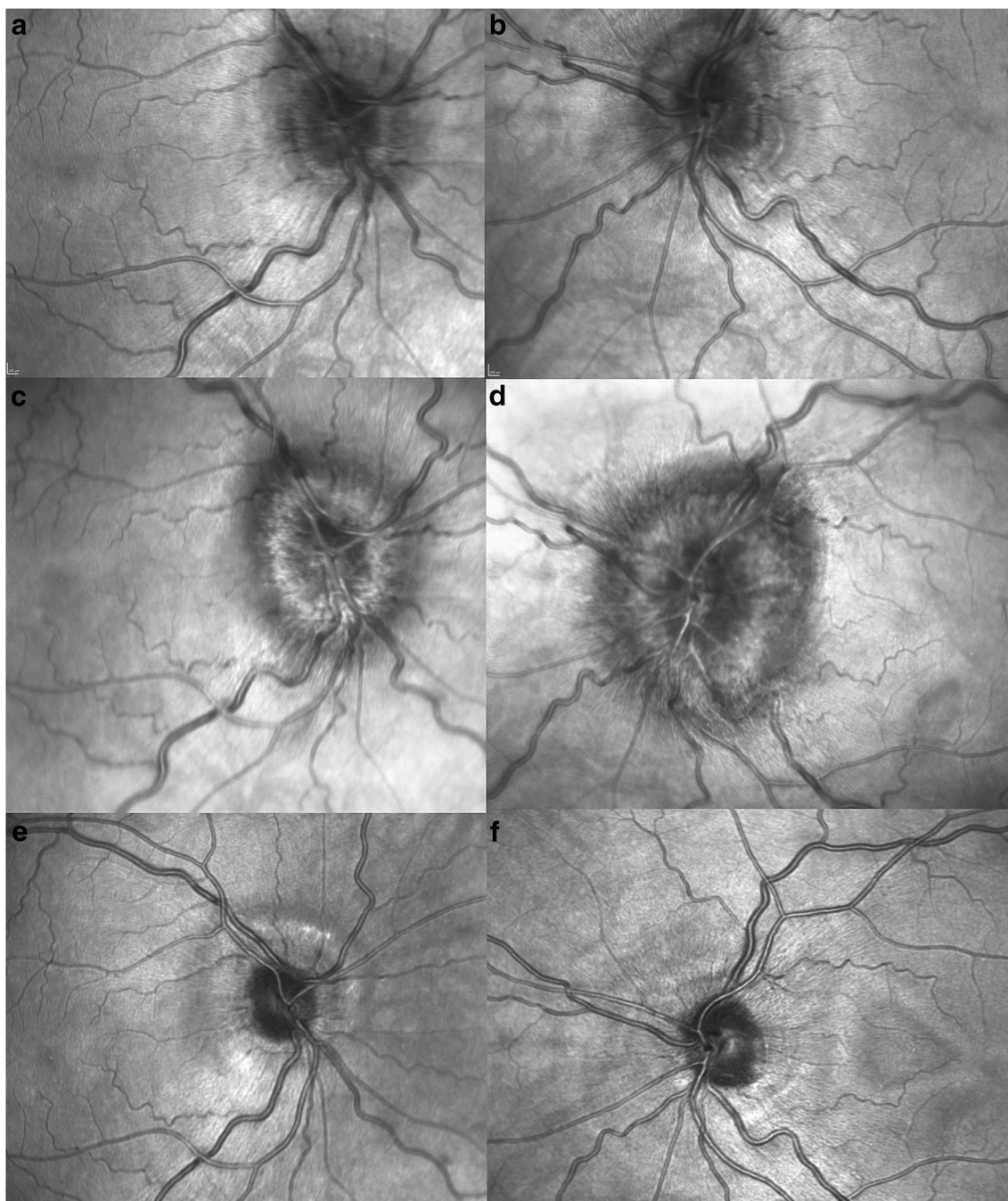
Fig. 1 Optical coherence tomography (OCT) imaging (Heidelberg Engineering, Heidelberg, Germany) infrared reflectance fundal images demonstrating changes in papilledema over the clinical course. **a** Right eye and **b** left eye at baseline; **c** right and **d** left at fulminant presentation; **e** right and **f** left 6 weeks post VP shunt with complete resolution of the papilledema

aim was to understand the disorder more comprehensively and to recognise when surgical intervention is warranted [15].

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 6 years. On presentation, her symptoms had returned with pulsatile tinnitus; daily headache; visual blurring; and transient visual obscurations on coughing or straining. Her headache fulfilled the criteria for "headache attributed to IIH" in the international classification of headache disorders 3; the character of the headache was chronic migraine-like, and it was daily with 8 migrainous
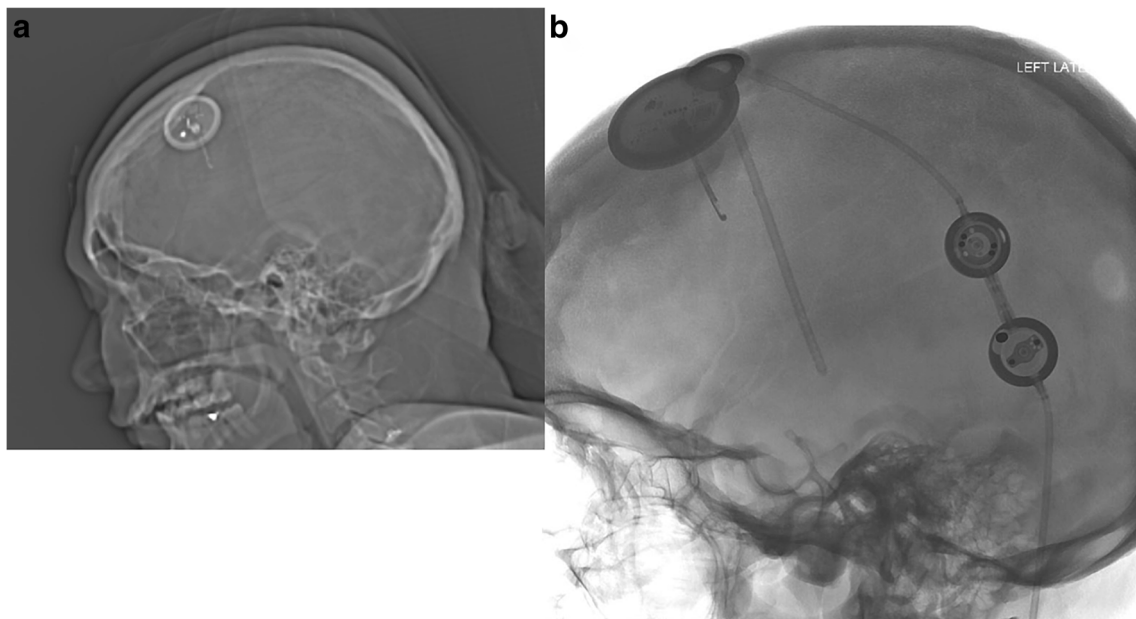
Fig. 2 **a** X-radiograph of skull showing telemetric Rau monitor. **b** X-radiograph of skull showing telemetric catheter and ventricular shunt placement

exacerbations per month with severe throbbing pain and nausea, photophobia, and phonophobia; she also described visual aura. 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 33 kg/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 35 cm CSF, with normal constituents. She had been started on topiramate 100 mg BD and acetazolamide 250 mg QDS prior to referral.

She was recruited to a research study; prior to enrolment, acetazolamide and topiramate were discontinued in line with the study protocol. Following enrolment, 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. The 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. 4c, 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 [6, 11]; whilst awaiting a definitive neurosurgical intervention, she was not treated with medication for raised ICP but did receive simple analgesia; no effect of medication on ICP was discerned. The trace, during the lumbar drain insertion, shows a marked drop of ICP at the point of insertion (Fig. 3c). The lumbar drain was clamped, 3 days after insertion, before the procedure to facilitate the insertion of the ventricular catheter in the slit ventricles. The lumbar drain had not reversed the underlying disease process and ICP rose from mean 2.0 mmHg (equivalent to 2.7 cm CSF) to mean 40.6 mmHg (equivalent to 55.2 cm CSF) over 2 h (Fig. 3d). The ICP postoperatively settled, with a mean of 18.9 mmHg (equivalent to 25.7cmCSF) (Fig. 3e).

Discussion

The case reports a 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 76 mmHg, with a mean ICP of 60 mmHg over a 10-min period (Fig. 3b). During these peaks, the patient reported a very severe headache on a verbal rating scale of 10. As the shunt insertion could not happen within 24 h, a lumbar drain was inserted, as recommended by the IIH guidelines [6, 11].

Fig. 3 Telemetric intracranial pressure monitoring in a case of fulminant IIH. **a** Baseline ICP trace, mean ICP 23.7 mmHg range 14.0–58.0 mmHg. **b** ICP trace during admission for fulminant IIH prior to intervention, mean ICP 33.3 mmHg, range 9.2–76.0 mmHg, period marked 'A' wave corresponded to a mean 76 mmHg, the highest maximal peak recorded during this period was 111.3 mmHg (not shown). **c** ICP trace during lumbar drain insertion (arrow), mean pre-41.5 mmHg range 15.4–70.6 mmHg, post insertion mean 14.7 mmHg, range 1.8–40.0 mmHg. **d** 2-h ICP trace recorded after closure of lumbar drain, initial mean 2.0 mmHg, shaded area mean 40.6 mmHg range 26.6–77.0 mmHg. **e** ICP trace recorded post-op ventricular-peritoneal shunt insertion, mean 18.9 mmHg, range 8.6–36.6 mmHg. All ICP monitoring shown was in the supine position, ICP trend data shown at 1 Hz, 1-h trace shown aside panel **d** 2 h. 1 mmHg converts to 1.36 cm H_2O. Mean and range automatically calculated by the software, DataView version, Raumedic, Helmbrechts, Germany

Time
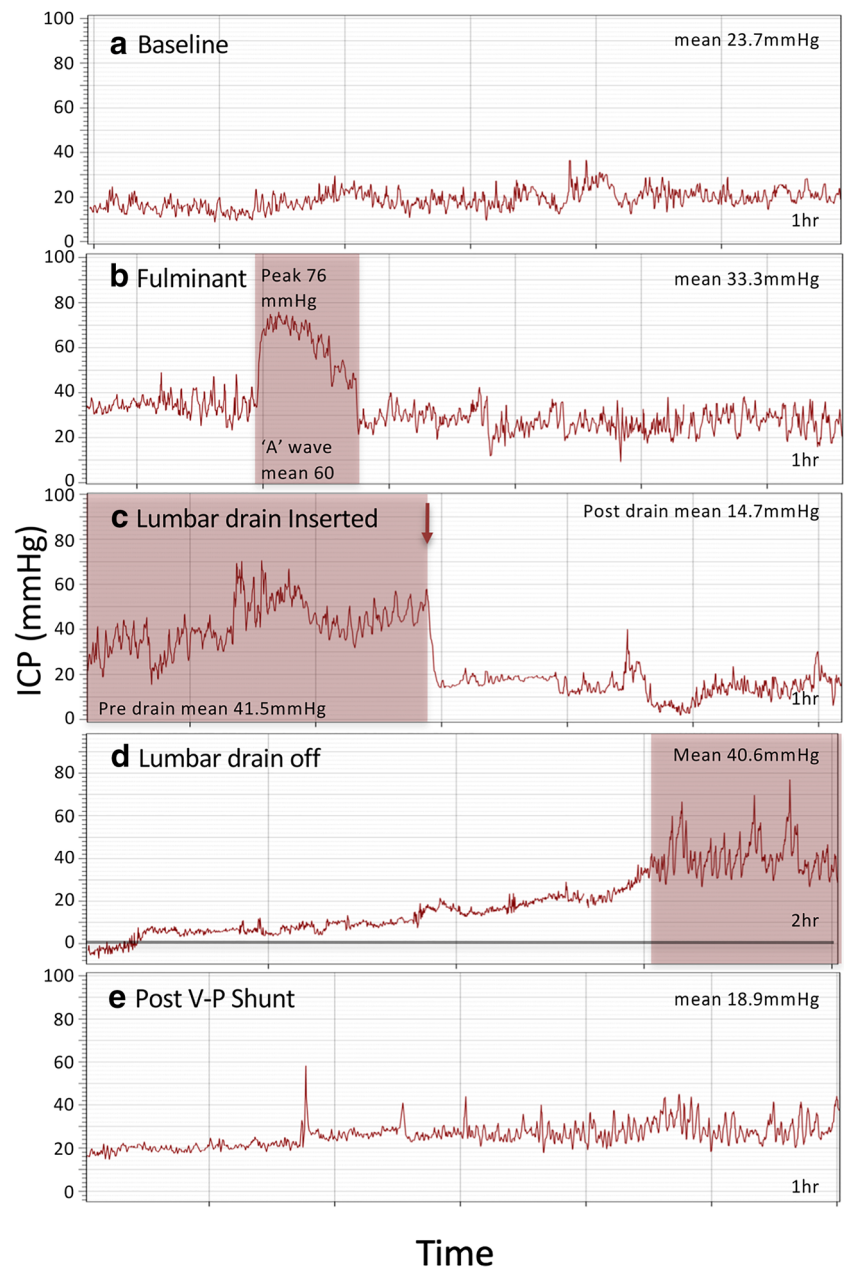
During this recording, we see a mean ICP of 14.7 mmHg (Fig. 3c), demonstrating the effectiveness of the drain in controlling ICP. Secondly, when the drain is clamped, just prior to surgery to effect filling of the ventricles, the ICP rapidly rises within 2 h to a mean of 40.6 mmHg (Fig. 3d). This last 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 [6, 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, 18, 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 [9]. Invasive measurements of ICP are often not warranted in this disease, as there is little evidence to suggest absolute pressure measures drive clinical decision-
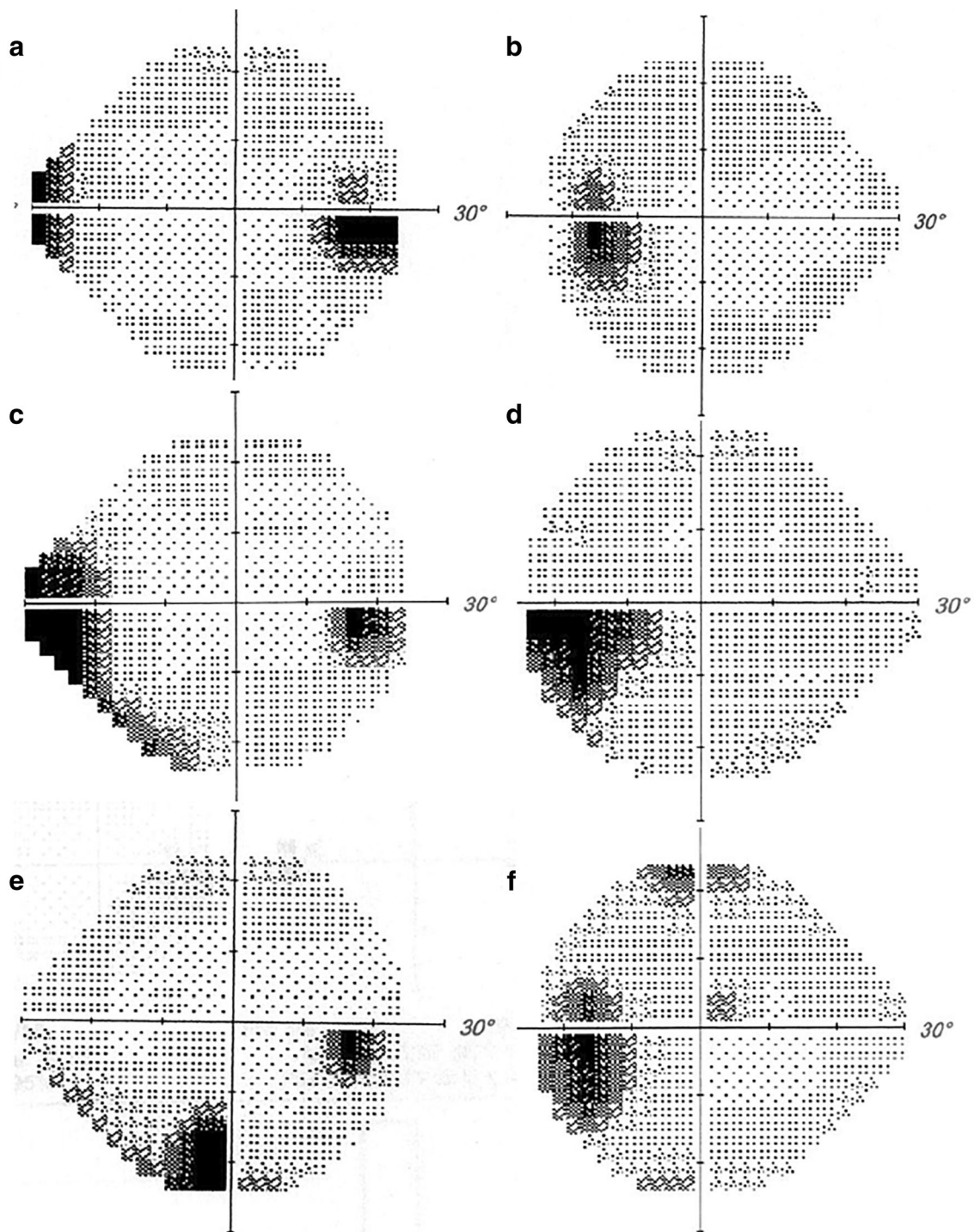
Fig. 4 Grey scale of the Humphrey visual fields. **a** Right eye at baseline showing enlarged blind spot and nasal step; **b** left eye at baseline showing enlarged blind spot; **c** right eye at fulminant presentation increase in the nasal step; **d** left eye at fulminant presentation increase in the 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

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
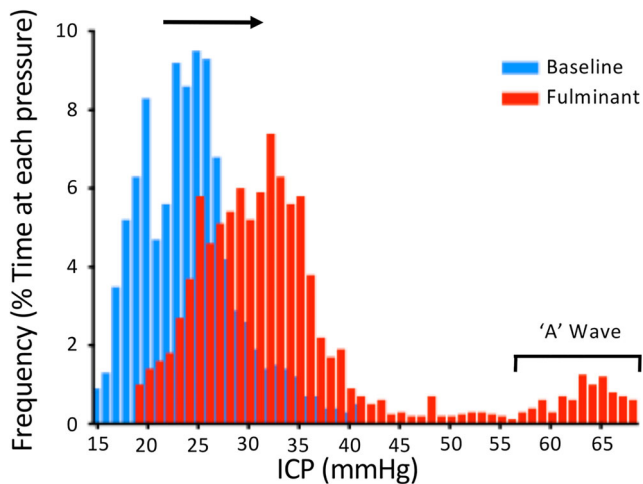
Fig. 5 Histogram showing ICP frequency at baseline and the shift in ICP observed with the fulminant presentation

seizures affecting 3% and infection in 1.5% [2]. It should however be noted that this was in a series of patients with significant structural brain abnormalities 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 a drift of 2.5 mmHg (3.2 cm H_2O) over a median of 8 months [17] and in some patients used for up to 5 years.

Telemetric ICP monitors have an evolving role in the diagnosis and monitoring of CSF disorders, both hypertensive and hypotensive. 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, low-pressure headache, and non-pressure-related headaches such as migrainous headaches or medication overuse headache [7, 8]. This is particularly important in IIH, where exacerbation of headache in the shunted patient can be complicated to investigate [12].

Conclusion

Telemetric ICP monitors are increasingly being used to manage difficult cases of IIH (although in this patient the monitor was originally cited 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.

Conflict of interest JLM—None.
SPM—Invex therapeutics, advisory board (2020).
GT—None.
ASJ—Invex therapeutics, company director with salary and stock options (2019, 2020).

References

1. Adderley NJ, Subramanian A, Nirantharakumar K, Yiangou A, Gokhale KM, Mollan SP, Sinclair AJ (2019) Association between idiopathic intracranial hypertension and risk of cardiovascular diseases in women in the United Kingdom. JAMA Neurol 76(9): 1088–1098

2. Antes S, Tschan CA, Kunze G, Ewert L, Zimmer A, Halfmann A, Oertel J (2014) Clinical and radiological findings in long-term intracranial pressure monitoring. Acta Neurochir (Wien) 156(5): 1009–1019

3. Antes S, Tschan CA, Heckelmann M, Breuskin D, Oertel J (2016) Telemetric intracranial pressure monitoring with the raumedicneurovent P-tel. World Neurosurg 91(C):133–148

4. Barber JM, Pringle CJ, Raffalli-Ebezant H, Pathmanaban O, Ramirez R, Kamaly-Asl ID (2016) Telemetric intra-cranial pressure monitoring: clinical and financial considerations. Br J Neurosurg 31(3):300–306

5. Duits FH, Lage PM, Paquet C et al (2016) Performance and complications of lumbar puncture in memory clinics: results of the multicenter lumbar puncture feasibility study. Alzheimers Dement 12(2):154–163

6. Hoffmann J, Mollan SP, Paemeleire K, Lampl C, Jensen RH, Sinclair AJ (2018) European Headache Federation guideline on idiopathic intracranial hypertension. J Headache Pain 19(1):982

7. Ishihara S, Fukui S, Otani N, Miyazawa T, Ohnuki A, Kato H, Tsuzuki N, Nawashiro H, Shima K (2009) Evaluation of spontaneous intracranial hypotension: assessment on ICP monitoring and radiological imaging. Br J Neurosurg 15(3):239–241

8. Lilja A, Andresen M, Hadi A, Christoffersen D, Juhler M (2014) Clinical experience with telemetric intracranial pressure monitoring in a Danish neurosurgical center. Clin Neurol Neurosurg 120:36–40

9. Mitchell JL, Mollan SP, Vijay V, Sinclair AJ (2019) Novel advances in monitoring and therapeutic approaches in idiopathic intracranial hypertension. Curr Opin Neurol 32(3):422–431

10. Mollan SP, Aguiar M, Evison F, Frew E, Sinclair AJ (2018) The expanding burden of idiopathic intracranial hypertension. Eye 81:1159

11. Mollan SP, Davies B, Silver NC et al (2018) Idiopathic intracranial hypertension: consensus guidelines on management. J Neurol Neurosurg Psychiatry 89(10):1088–1100

12. Mollan SP, Hornby C, Mitchell J, Sinclair AJ (2018) Evaluation and management of adult idiopathic intracranial hypertension. Pract Neurol 18(6):485–488

13. Mollan SP, Hoffmann J, Sinclair AJ (2019) Advances in the understanding of headache in idiopathic intracranial hypertension. Curr Opin Neurol 32(1):92–98

14. Mollan SP, Mitchell JL, Sinclair AJ (2019) Tip of the iceberg in idiopathic intracranial hypertension. Pract Neurol 19(2):178–179

15. Mollan S, Hemmings K, Herd CP, Denton A, Williamson S, Sinclair AJ (2019) What are the research priorities for idiopathic intracranial hypertension? A priority setting partnership between patients and healthcare professionals. BMJ Open 9(3):e026573

16. Mulla Y, Markey KA, Woolley RL, Patel S, Mollan SP, Sinclair AJ (2015) Headache determines quality of life in idiopathic intracranial hypertension. J Headache Pain 16(1):461

17. Norager NH, Lilja-Cyron A, Bjarkam CR, Duus S, Juhler M (2018) Telemetry in intracranial pressure monitoring: sensor survival and drift. Acta Neurochir (Wien) 121(4):797–798

18. Scotton WJ, Mollan SP, Walters T, Doughty S, Botfield H, Markey K, Yiangou A, Williamson S, Sinclair AJ (2018) Characterising the patient experience of diagnostic lumbar puncture in idiopathic intracranial hypertension: a cross-sectional online survey. BMJ Open 8(5):e020445

19. Xu DS, Hlubek RJ, Mulholland CB, Knievel KL, Smith KA, Nakaji P (2017) Use of intracranial pressure monitoring frequently refutes diagnosis of idiopathic intracranial hypertension. World Neurosurg 104:167–170

20. Yiangou A, Mitchell J, Markey KA, Scotton W, Nightingale P, Botfield H, Ottridge R, Mollan SP, Sinclair AJ (2018) Therapeutic lumbar puncture for headache in idiopathic intracranial hypertension: Minimal gain, is it worth the pain? Cephalalgia 39(2):245–253