

Letter: Cerebrospinal fluid shunting for idiopathic intracranial hypertension

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1 To the Editor:

2 Salih et al¹ recently published a meta-analysis on cerebrospinal fluid (CSF) shunting in
3 Idiopathic Intracranial Hypertension (IIH). This publication amongst others² are important in
4 highlighting the lack of detail provided in common data points and the need for uniformity in
5 reporting outcome measures when assessing the success of any intervention. IIH is a condition
6 managed by neurologists, ophthalmologists, neurosurgeons and more recently interventional
7 radiologists. Each specialist sees a different angle of the condition however the contemporary
8 literature is there to guide us in the advances in our understanding of the disease and how to
9 manage it.

10

11 IIH is a systemic metabolic disease and comes under the classification of pseudotumour cerebri
12 syndrome (PTSC).³ This distinction is important as PTCS may be primary (IIH) or arise from an
13 identifiable secondary cause such as anaemia, medication use or indeed cerebral venous sinus
14 thrombosis (CVST).³⁻⁵ Outcomes from shunting in secondary pseudotumor versus IIH can be
15 expectantly different, and why classifying the underlying pathology in this context is essential.
16 The authors have not detailed this nuance for readers nor provided an assessment of quality of
17 the studies specifically against any version of the diagnostic criteria. The 2002 Friedman and
18 Jacobson criteria⁶ are cited as useful for diagnosis, which is in part true but for the time frame
19 when the majority of the included studies performed data capture. However in modern practice
20 these have been superseded by the revised diagnostic criteria for PTSC which were published in
21 2013.³

22

23 A key concern we have is the discussion detailing management strategies which was not the
24 focus of the meta-analysis. Papilloedema is a key single point of entry to the investigational
25 pathway,³⁻⁵ which has been omitted in their discussion. We agree that establishing the diagnosis
26 of IIH early is important but this is not based on symptoms of headache, visual loss or visual
27 changes and tinnitus, which are nonspecific. Yri and Jensen⁷ eloquently showed that in a control
28 population one third reported transient visual obscurations, one quarter had pulsatile tinnitus, and
29 one quarter experienced double vision. Indeed the controls' headache phenotype had features of
30 a raised ICP headache. More intriguing still was that one quarter of those without IIH had

31 improvement of their headache following lumbar puncture. We, and a multidisciplinary group,
32 therefore strongly discourage the use of symptoms to guide surgical management in IIH.^{4,5}

33

34 We agree that venography is absolutely critical in the assessment of papilledema to exclude
35 CVST which has devastating consequences if missed.^{4,5} We would be concerned about this over
36 emphasis on DVSS in the absence of randomized control trial evidence. The tsunami of
37 institutional based case series in stenting is challenging to unpick the indications for the
38 intervention and indeed the heterogenous outcomes that have been reported.⁸ We agree there is a
39 role for DVSS in IIH and to address this gap in the literature we are spearheading a randomized
40 control trial in the United Kingdom for the evaluation of CSF diversion and DVSS in those with
41 visual failure in IIH.

42

43 We are encouraged by the reducing complication rate reported.¹ This too has been our local
44 experience which we believe is due to a dedicated CSF shunt pathway, CSF specialist surgeons,
45 and a standardized protocol: where there is a preference for a frontal ventriculoperitoneal shunt;
46 use of adjustable gravitational valves; an implantable ICP monitoring device in line with the
47 shunt to optimize CSF drainage and identify possible future malfunction; frameless stereotactic
48 image-guidance for insertion of the ventricular catheter; and laparoscopic placement of the
49 peritoneal end in patients with high body mass index.^{9,10}

50

51 Horizon scanning in IIH reveals an avenue where the need for surgical intervention could be
52 reduced. This has been the translation of Glucagon-like peptide-1 receptor (GLP-1R) agonists
53 from an animal model with raised ICP (hydrocephalic)¹¹ to the first phase 2 study assessing the
54 biological effect of the GLP-1R agonist Exenatide on ICP utilising highly accurate telemetric
55 ICP monitors. Exentide was successful in reducing ICP at 2.5hours, 24 hours and 12 weeks.¹²

56

57 To our final point there is firm evidence that IIH is a metabolic disease encompassing a spectrum
58 of systemic manifestations. There is twofold risk of cardiovascular disease, presence of insulin
59 resistance, increased truncal adiposity, hyperleptinemia, fertility and pregnancy complications.¹³⁻

60 ¹⁵ These features are in excess than that driven by the presence of obesity. Adipose tissue in IIH
61 has a unique profile of transcription and metabolic dysregulation driving lipogenesis and

62 promoting increased adipose deposition.¹⁵ Hormonal perturbations have been identified with
63 patients noted to have a unique phenotype of serum and CSF androgen excess and a dysregulated
64 glucocorticoid phenotype.^{16,17} Weight loss is now established as the only disease modifying
65 therapy with bariatric surgery producing long term control of ICP.¹⁸ While venous sinus stenosis
66 is almost universally observed in active IIH, we strongly believe this reflects a consequence of
67 the disease rather than the casual driver in the majority.

68

69 **Keywords:** Cerebrospinal fluid shunt, Dural Venous Sinus Stenting, Headache, Idiopathic
70 Intracranial Hypertension, ICP, Papilloedema, Pseudotumour cerebri.

71

72 **Abbreviations:** CSF = cerebrospinal fluid; DVSS = dural venous sinus stenting; ICP =
73 intracranial pressure; IIH = idiopathic intracranial hypertension;

74

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