Ophthalmological Detection of Intracranial Hypertension in Craniosynostosis: A Diagnostic Accuracy Study

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Introduction: Craniosynostosis is characterised by the premature fusion of cranial sutures. It is often associated with intracranial hypertension (IH) and can cause cognitive impairment, visual impairment and death, if untreated.[1] Invasive intracranial pressure (ICP) measurement represents the gold standard, but requires hospital admission and carries risk.[2] Here, we have applied ophthalmological methods to non-invasively detect IH in craniosynostosis.

Aim: To assess the diagnostic accuracy of fundoscopy and visual evoked potentials (VEPs) in detecting IH in patients with craniosynostosis.

Methods:

Single-centre, retrospective diagnostic accuracy study

Inclusion criteria:

- Children with craniosynostosis undergoing 48-hour ICP and surgery

Main outcome measures:

- Primary: fundoscopy, VEPs and IH, defined as ICP>20 mmHg
- Secondary: Final VA

Results: Median age at first presentation was 3.8 months (range: 0.1-59.1); median follow-up was 80.9 months (range: 18.5-156.2). Figure 1 displays the patient inclusion flowchart. Figure 2 displays diagnostic accuracy results. Median final VA was 0.24 logMAR (range: -0.06 to 2.7); 26 patients (72.2%) achieved UK driving standard VA, defined as at least 6/12 in the better eye.

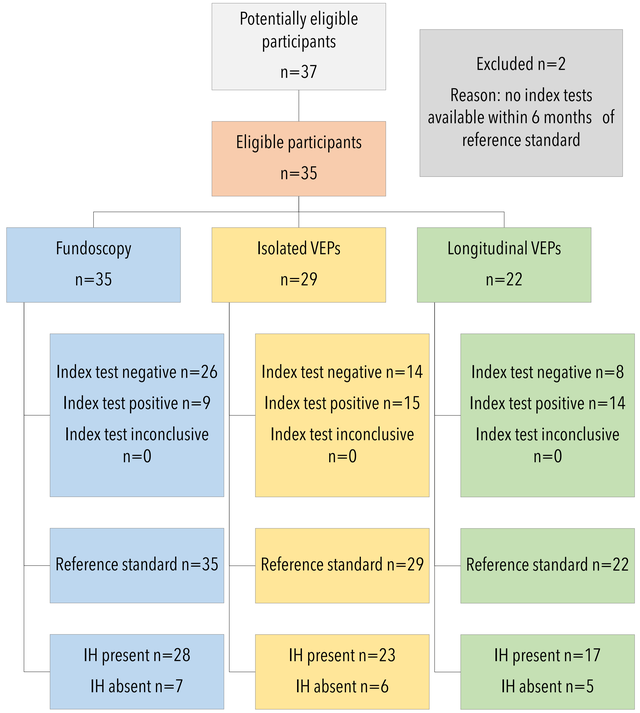


Figure 1: Patient inclusion flowchart

Isolated VEPs recorded within 6 months of ICP assessment; Longitudinal VEPs recorded as per isolated, plus two preceding visits; IH = intracranial hypertension; VEPs = visual evoked potentials.

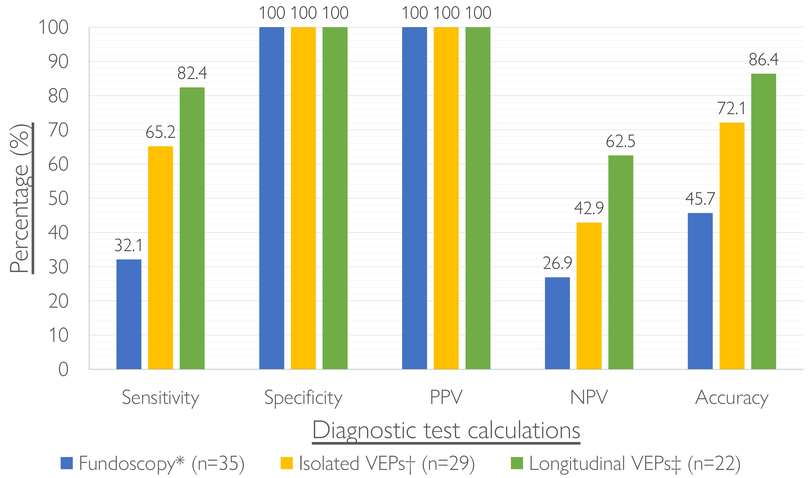


Figure 2: Diagnostic accuracy results

**ICP assessments and fundoscopy within 6 months of ICP assessments available for 35 children. †Isolated VEPs recorded within 6 months of ICP assessment. ‡Longitudinal VEPs recorded as per isolated, plus two preceding visits. Accuracy = true test results / all evaluated cases; NPV = negative predictive value; PPV = positive predictive value.*

Conclusions: Papilloedema present on fundoscopy reliably indicated IH, but its absence did not exclude IH (consistent with Tuite *et al*).[3] VEP monitoring demonstrated higher sensitivity for detecting IH, with serial testing increasing sensitivity even further.

References: 1. Renier D, et al. J Neurosurg. 1982 Sep;57(3):370-7.
2. Tamburrini G, et al. Childs Nerv Sys. 2005 Oct;21(10):913-21.
3. Tuite GF, et al. Neurosurgery. 1996 Feb;38(2):272-8.

Competing interests: None. **Funding:** Sohaib Rufai is funded by a National Institute for Health Research (NIHR) Doctoral Fellowship for this research project. This poster presents independent research funded by the National Institute for Health Research (NIHR). The views expressed are those of the author(s) and not necessarily those of the NHS, the NIHR or the Department of Health and Social Care. **Correspondence:** Sohaib.Rufai@nhs.net