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 followup 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.

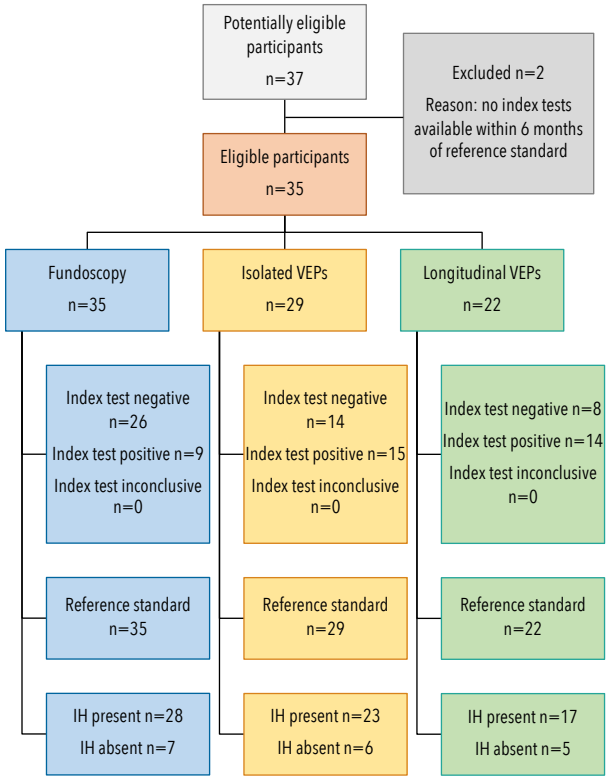

Figure 1: Patient inclusion flowchart

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.

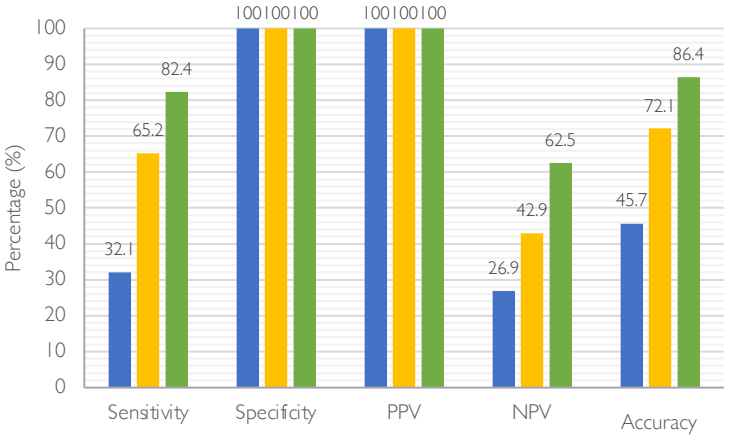

Figure 2: Diagnostic accuracy results

**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.

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