Letters to the Editor

NEUROSURGICAL FORUM

Raised intracranial pressure and nonsyndromic sagittal craniosynostosis

TO THE EDITOR: The recent paper by Wall et al.19 (Wall SA, Thomas GPL, Johnson D, et al: The preoperative incidence of raised intracranial pressure in nonsyndromic sagittal craniosynostosis is underestimated in the literature. J Neurosurg Pediatr 14:674–681, December 2014) on intracranial pressure (ICP) in a selected group of untreated children (mean age around 5 years) with sagittal synostosis ends with the recommendation that intraparenchymal ICP monitoring “should be considered and used routinely in all patients for whom a nonoperative course of management is proposed, no matter the nature or severity of the calvarial deformity.” As this represents a radical departure from the clinical practice of many, if not most, craniofacial units and because we fear the recommendation could lead to an epidemic of invasive monitoring (with its small but never absent risk of brain injury), we suggest that the following factors be considered.

Most importantly, their recommendation is associated with no identified benefit to the patients in their study. Not only do they state that “developmental delay was not significantly more common in patients with elevated ICP,” but they also reveal that “patients who manifested symptoms suggestive of raised ICP but who were found to have normal ICP on monitoring experienced an improvement or resolution of their symptoms over time.” There is no mention of what happened to those who did have abnormal ICP. In fact, the only support for their recommendation is their statement, “It remains reasonable to assume that continued chronically raised ICP in [sagittal craniosynostosis (SC)] is liable to curtail an individual’s neurocognitive development, even if it does not reliably cause a marked developmental delay and is not responsible for many of the developmental abnormalities seen in SC.” But is such an assumption in the absence of any supporting evidence “reasonable” enough to bear responsibility for one, possibly two, surgical interventions—one of which is a major cranial procedure?

Then there is the problem of what childhood ICP should be considered abnormal. The “true” level of childhood ICP remains, for obvious reasons, unknown. As Dominique Renier and his colleagues wrote in their classic 1982 paper,13 “The definition of ‘normal’ and ‘abnormal’ ICP recordings in children raises an initial problem.” Although the “Paris” range13 (< 10 mm Hg normal, 11–15 mm Hg borderline, and > 15 mm Hg raised) used here is most frequently accepted in craniofacial circles, a close reading of the 3 references5,9,17 used to support those values suggests the upper limit of their range could just as reasonably have been raised to 20 mm Hg.

Indeed, in a previous paper8 from Wall and colleagues’ unit (similar in its message but dealing with unicoronal synostosis), the authors state that “intracranial pressure was defined as elevated if the mean pressure was greater than or equal to 20 mm Hg.” It would be interesting to know their reasons for a change which, as they state in their Discussion, would reduce the number with raised ICP in their 39 monitored patients from 17 (44%) to 6 (15%)—and that’s without the 6 who were not monitored at all but were “well and asymptomatic when last reviewed.”

Their inclusion of “more than 3 B-type waves in a 24-hour period during sleep” as part of their classification of abnormal ICP is not helpful in the absence of any accompanying definition. “B” waves have had a checkered history since Lundberg first identified them in a study dealing predominantly with intracranial tumors in adults. Eide et al. pointed out9 (in a paper referenced by Wall et al.) that “the identification of B waves is subjective” and for that reason did not consider them useful in the diagnosis of raised ICP. In Renier et al.’s 1982 study13 waves superimposed on a non-elevated baseline ICP (their “third type”) were dismissed as normal. In Wall et al.’s study,9 all 6 patients with 3 or more B-waves had baseline pressures of or below 15 mm Hg. Nevertheless, their presence was judged sufficiently abnormal to justify calvarial remodeling (surgical details not given despite some techniques producing a temporary rise in ICP).

The waves of increased ICP superimposed on an elevated baseline observed in children with syndromic/complex forms of craniosynostosis occur particularly during REM sleep—associated episodes of airway compromise. Although children with single suture synostosis are unlikely to be so affected, it does raise the question of what effect a “normal” degree of childhood adenotonsillar hypertrophy may have on ICP. To take an extreme example, it could be argued that any child (with or without craniosynostosis) who snores at night runs an ICP that, according to the criteria quoted in the authors’ study, would qualify them for cranial vault surgery. On a lighter note, the authors’ observation that raised ICP was seen less frequently in children with obvious scaphocephaly than in those in whom it was minimal or absent altogether might reflect no more than a tendency for the former to sleep more comfortably on their sides than their backs (when a vulnerable airway may be compromised).
There is indeed a small but definite incidence of papilledema-confirmed increased ICP in children with isolated sagittal synostosis. Its cause is unknown, but in the absence of other remediable factors (hydrocephalus, for example), it represents a proper indication for surgical intervention—usually a procedure designed to increase the cranial volume. None of the patients reported here had papilledema.

The authors state that “the sequelae of chronically raised ICP include visual loss and psychomotor impairment.” Visual loss due to papilledema—which, again, none of these patients had—is not in dispute. But with regard to possible brain effects, neither of their supporting references do more than repeat that statement. Pollack et al. reference Renier and colleagues (plus a 1988 Paris review, which added further numbers to the earlier paper) as their authority, whereas Vinchon et al. quote Connolly et al., who also do no more than refer back to Renier’s 1982 paper with its various pitfalls.

Given that only 6 patients had postoperative ICP monitoring, whether surgery actually lowered ICP in the remainder remains a matter of conjecture.

The authors quote Hanlo et al. in support of their statement that “in other pediatric neurological conditions, elevated ICP has been correlated with impaired myelination and later poor neurodevelopmental scores.” But since the authors of that study looked at a condition (progressive hydrocephalus in infancy) with a very different mechanism for brain injury compared to craniosynostosis (and relied on anterior fontanelle tonometry to assess ICP), its relevance here is limited.

Finally, the authors quote 3 references in their statement that “several PET studies in children with isolated single-suture craniosynostosis have found areas of cerebral hypoperfusion associated with the stenosed suture, which resolved after corrective surgery.” In fact, only one of those references deals with PET scanning, and the other two refer to SPECT. A recent PubMed search failed to produce any other relevant PET studies.

In conclusion, we do not doubt that the content of Wall et al.’s paper is, in purely observational terms, correct; selected children with untreated sagittal synostosis of variable severity monitored at a mean age of around 5 years may be running ICPs at or above 15 mm Hg. What this means in terms of their cognitive development, however, is quite another matter. As the authors state, “The lack of a correlation between developmental delay and raised ICP found in the present study supports the hypothesis that there is no simple causal relationship between ICP and neurocognitive anomalies found in SC.” This leaves their recommendation that, in pursuit of essentially theoretical neurocognitive benefits, invasive ICP monitoring with a view to “calvarial remodeling” (with the attendant brain risks of both procedures) should be performed in all children whose parents have declined surgical intervention at least debateable—at worst dangerous.

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DISCLOSURE
The authors report no conflict of interest.

References