Neurosurgical Forum LETTERS TO THE EDITOR

Raised intracranial pressure and cognitive delay in craniosynostosis

TO THE EDITOR: In their perspective piece published in the Broca's Area section of *JNS: Pediatrics*, Hayward et al.⁶ describe and debate a lack of causal evidence linking the finding of raised intracranial pressure (ICP) and neurocognitive impairment (NCI) in children with syndromic craniosynostosis (Hayward R, Britto J, Dunaway D, et al: Connecting raised intracranial pressure and cognitive delay in craniosynostosis: many assumptions, little evidence. *J Neurosurg Pediatr* 18:242–250, August 2016), the implication being that raised ICP may not be the major contributor to NCI. Our thoughts on this matter, and the consequences for screening and treatment of these children, are discussed below.

Causality Between Raised ICP and NCI in Syndromic Craniosynostosis

Table 1 summarizes the findings in craniosynostosis due to various genetic causes. Of note, the rate of raised ICP appears to be related to the underlying genetic abnormality: a high rate for *FGFR2*-related craniosynostosis (Apert and Crouzon/Pfeiffer syndromes); a low rate for *P250R FGFR3* craniosynostosis (Muenke syndrome); and an intermediate rate for *TWIST1*-related craniosynostosis (Saethre-Chotzen syndrome). The intelligence quotient (IQ) data reported for each of these groups show either no obvious correlation between raised ICP and IQ or that such a correlation is concealed by a more major impact of the underlying genetic defect. For example, in Muenke syndrome there is low prevalence of raised ICP, but such

individuals are twice as likely to have an IQ less than 85. In contrast, in Crouzon syndrome there is high rate of raised ICP but in the context of near-normal IQ for this population.

Causes of Intracranial Hypertension

In our opinion, once raised ICP is detected, subsequent treatment should be aimed at its normalization. To establish any effect of prolonged raised ICP on IQ would require not only a deviation from our usual practice but also a prolonged period of observation in which patients would be unnecessarily exposed to the risk of visual loss.

We prefer to use the term “intracranial hypertension” (ICHT) instead of the term “raised ICP” to better reflect the totality of an unwanted condition or state in syndromic craniosynostosis. Four factors contribute to ICHT. First, Spruijt et al.¹³ showed that skull growth impairment using the occipitofrontal head circumference (OFC), particularly by the age of 3 years, was a big contributor to the development of ICHT. The authors described this finding in 13 of 20 syndromic craniosynostosis patients within their first 6 years of life. That is, even though the intracranial volume is higher than normal in syndromic craniosynostosis,³ the cessation of growth has significant effects. The second contributor to ICHT is obstructive sleep apnea (OSA).⁵ In our recent study, moderate and severe OSA were associated with ICHT while mild OSA was not.¹³ Next, there is the presence of hydrocephalus. Although hydrocephalus has low prevalence in syndromic craniosynostosis, when it occurs it is most closely related to Crouzon syndrome.¹ Commonly, patients present with a nonprogressive form of ventricular enlargement (i.e., ventriculomegaly). In general, this finding does not require treatment, but this practice

TABLE 1. Intracranial pressure, IQ, and the presence of venous collaterals in syndromic craniosynostosis and in controls

Syndrome or Grouping	Rate of Raised ICP*	Full-Scale IQ†		Percentage w/ Full-Scale IQ <85†	Occipital Collaterals Score, Mean‡	All Collaterals Score‡	
		Mean	Range			Mean	Range
Apert	50%	77	59–94	67%	0.55	11.4	5–17
Crouzon/Pfeiffer	55%	103	54–133	22%	0.33	11.3	7–17
Muenke	11%	95	73–124	39%	0	10.7	7–15
Saethre-Chotzen	33%	100	52–141	21%	0.33	14.0	11–16
Controls	Very low	100	47–188	16%	0	2.0	—

* Prevalence of papilledema during the first 6 years of life. Data from Spruijt et al.¹³

† Full-scale IQ determined in children aged 6–13 years with genetically confirmed diagnoses. Data from Maliepaard et al.⁷

‡ Presence of venous collaterals on CT angiogram; occipital collaterals score varies from 0 to 2, and all collaterals score from 0 to 22. Data from Florissson et al.⁴

has not been substantiated scientifically. Last, we should consider the contribution to ICHT made by obstruction to venous outflow from the cranial vault.

As mentioned by Hayward et al.,⁶ venous outflow obstruction is difficult to quantify since we only have indirect studies available to us. That said, venous outflow obstruction appears to be a common feature in syndromic craniosynostosis patients with ICHT, but it is not regarded as a likely contributor to NCI. In our transcranial Doppler ultrasound (TCD) study of 12 children with syndromic craniosynostosis (before vault expansion and without hydrocephalus), we presented indirect evidence that would be consistent with venous outflow obstruction. For example, the mean middle cerebral artery (MCA) cerebral blood flow velocity (CBFv), peak systolic CBFv, end-diastolic CBFv, resistive index, and systemic blood pressure were all higher in the 6 patients with ICHT than in those without ICHT.¹⁵ (The abnormal resistive index reflects cerebrovascular outflow resistance beyond the MCA and anywhere in the microcirculation, and more distally.) Three weeks and 3 months after vault expansion, all TCD values were similar for both groups, but the difference in systemic blood pressure remained. More directly focusing on the anatomy of cerebral venous outflow, Rich et al.¹¹ described anomalies in the occipital venous drainage system. In our experience, reported by Florisson et al.,⁴ all craniosynostosis patients had a smaller diameter of the jugular foramen than normal controls, while in 15% of cases the jugular vein did not pass through the jugular foramen. Occipital collateral veins were present particularly in *FGFR2*- and *TWIST1*-associated craniosynostosis and not in P250R *FGFR3* craniosynostosis (Table 1). These collaterals are already present at a very young age and independent of the presence of subsequent ICHT.

In regard to the development of the cranial venous system, it is also apparent that the number of collaterals remains stable over time. Therefore, we should consider this contribution to ICHT as an inborn developmental anomaly, due to the genetics. In an embryologic description of normal development of the cranial venous system, Padgett¹⁰ described the emergence of emissary veins, starting with the hypoglossal in Stage 4 (10–16 mm), the condylar in Stage 5 (16–21 mm), and the mastoid in Stage 6 (18–26 mm). At Stage 7 (2.5 months, 40 mm) the chondrocranium develops around the stem of the emissaries, thus forming their foramina. The later fetal period of development is also well described:⁹ until 4.5 months (17th and 18th week) the transverse sinus has a relatively even caliber, and it then enlarges at the lateral borders; within the next 1–1.5 months this ballooning reaches the midline; and next, the cortical veins start to drain into the transverse sinus, thereby increasing cerebral blood volume and venous drainage. The inner diameters of the sigmoid and jugular sinuses remain extremely small during intrauterine life. This feature combined with the extra blood flow in the transverse sinus causes a physiological cerebral venous hypertension, which induces the development of collaterals. The jugular sinus (later to be the jugular bulb) is surrounded by cartilage and bone, which might make it difficult to expand; this development needs the pulsating effect of ascending negative pulse waves from the right

atrium, present after assuming the erect position. After birth, the diameters of the sigmoid and jugular sinuses expand rapidly and the jugular bulb develops. In syndromic craniosynostosis the altered *FGFR2* or *TWIST1* gene appears to cause a cessation of normal development of the internal jugular vein, on one or both sides, and a reduced dimension of the jugular foramen.⁴ These collaterals enable a normal venous outflow most of the time in patients with syndromic craniosynostosis. If the collaterals are sacrificed during surgery, or when other factors compromise venous outflow (e.g., impaired skull growth, ventriculomegaly or hydrocephalus, or tonsillar herniation), or when blood inflow increases (e.g., during rapid eye movement [REM] sleep and apneas or hypopneas), venous hypertension occurs and results in ICHT.

Functional Impact of ICHT

Recently, we have also reported the impact of ICHT and/or OSA on sleep architecture. Patients with syndromic craniosynostosis who lacked signs of ICHT and/or OSA had normal sleep patterns. During sleep, ICHT on its own is associated with an increased rate of arousal. Moderate or severe OSA (with or without ICHT) results in severe disruption of sleep, with less REM sleep, more arousals, and reduced sleep efficiency.¹⁴ These findings indicate that besides OSA, ICHT by itself is associated with disrupted sleep patterns and thus may affect school achievement. In the same report,¹⁴ there are 5 patients in whom the sleep pattern normalized after monobloc advancement; since this procedure both treats OSA and expands intracranial volume, we cannot tell which of these factors contributes most to the improvement in sleep. Measurements of the quality of sleep may become useful in screening for ICP/ICHT, since these are objective and independent of the genetic cause of craniosynostosis. Better sleep may also be related to functional outcome and IQ testing results.

Treatment Strategy for Syndromic Craniosynostosis

Given the above discussion, we do not subscribe to the notion that unnecessary surgeries are being undertaken in patients with syndromic craniosynostosis. Since there is a low prevalence of ICHT in Muenke syndrome, one could argue that in this particular condition a strict wait-and-see policy could be considered. In Apert, Crouzon/Pfeiffer, and Saethre-Chotzen syndromes, the prevalence of ICHT is much higher. A wait-and-see policy before any surgery in these groups would require strict compliance with follow-up and adherence to use of a validated ICHT screening tool with excellent sensitivity and specificity. It is our belief that such a tool is not yet available; funduscopy in our center is a defining test for the presence of papilledema, but papilledema may still be absent despite elevated ICP. Optical coherence tomography (OCT) is a more objective tool than funduscopy, but it requires cooperation of the patient and is only feasible in patients 3–4 years of age. Assessment of electrophysiological visual evoked potentials is labor intensive and therefore not suitable for repeated screening in large numbers of patients. A simple and reliable measure that we routinely take at every visit—in addition to performing funduscopy—is the

OFC, since it is a good measure of intracranial volume in early childhood¹² and predicts intracranial hypertension.¹³ The combination of these assessments may improve the detection of ICHT, but the sensitivity and specificity of this approach are unknown.

The best way to determine whether routine early vault expansion is to be preferred over a wait-and-see policy can only be demonstrated by comparing the outcome of large cohorts of syndromic craniosynostosis patients. The outcome should include assessment of vision and neurodevelopment, the presence of tonsillar herniation, and neurological testing.

Our treatment protocol with early vault expansion for syndromic craniosynostosis is not based solely on the identification of raised ICP. We take a broader view of ICHT and in this context skull expansion surgery not only treats the problem of raised ICP but also other contributory components (see above). An alternative term that might better address our all-inclusive perspective is “intracranial compartment syndrome.” Despite the difference in philosophy between our center and that of Hayward and colleagues, the results are comparable in regard to the number of procedures performed.

Lastly, Hayward et al.⁶ also discuss the phenomenon of raised ICP in relation to single-suture synostosis. In our view, this topic requires a separate debate. On review of our cohort of 262 patients with metopic synostosis, the rate of ICHT (i.e., detected through routine fundoscopy, OFC measures, or OCT) was 1.9% before surgery (mean age 11 months, SD 2 months) and 1.5% (mean age at last follow-up 4.9 years) on follow-up.² Repeat operation for ICHT was performed in 2 patients (0.8%). The rate of raised ICP was 10.3% for sagittal synostosis, when surgery was delayed until the age of 11 months. The subsequent rate of raised ICP during an identical period of follow-up was highly dependent on treatment: 2.4% following early spring-mediated expansion, 6.8% following late frontobiparietal remodeling, and 8.9% following early biparietal outfracturing. The occurrence of ICHT in sagittal synostosis appears to be related to the intracranial volume achieved and the type and timing of surgery. But perhaps other factors that have been given little attention so far are also relevant, such as distortion of the brain. For example, according to Poiseuille's equation—

$$F \propto \frac{\Delta P \cdot r^4}{\eta \cdot L}$$

where F is flow, ΔP is perfusion pressure, r is vessel radius, η is blood viscosity, and L is vessel length—blood flow is vulnerable in regions supplied by long arterioles such as the centrum semiovale.⁸ This anatomical characteristic may have implications for cerebral blood flow in the severe scaphocephalic head shape and turriccephaly.

In conclusion, there is more to craniosynostosis than just ICP. More insight in the total system compliance of the intracranial compartment is required to truly understand normal physiology and its aberrations in craniosynostosis, followed by understanding the impact of interventions that we undertake.

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Disclosures

The authors report no conflict of interest.

Response

We are grateful to our friend Professor Mathijssen and her Rotterdam colleagues for adding their thoughts to our paper examining the connection between raised ICP and NCI in children with craniosynostosis. We particularly commend their succinct discussion of the genesis of the venous hypertension now recognized as such an important contributor to the elevation of ICP seen particularly in complex/syndromic forms.

The aim of our paper was also to stimulate discussion in regarding the question of whether some form of vault expansion should be the default approach to all children presenting with syndromic forms of craniosynostosis and to suggest that such linkage as might exist becomes attenuated with age.

We are delighted therefore that Professor Mathijssen and her colleagues agree that for patients with conditions with a low prevalence of raised ICP diagnoses, such as Muenke syndrome, “one could argue that ... a strict wait-and-see policy could be considered.” But is the neurocognitive development of children managed according to the more expectant policy we follow being put in jeopardy? Are they (as is suggested) being “unnecessarily exposed to the risk of visual loss”? Our experience of 49 children with Crouzon syndrome¹ suggests not. Under a default policy of cranial vault expansion at around 1 year of age, 19 children would have been subjected to an unnecessary, major cranial procedure, and, given that the mean age of a first episode of raised ICP was 1 year 2 months, it would be difficult to argue there had been material delay for the 16 (of 30) for whom investigations confirmed vault expansion to have been the appropriate treatment.

Professor Mathijssen and colleagues are surely right, however, to suggest that the association between raised ICP and NCI in single-suture synostosis requires detailed attention in its own right—an area in which they have opened a door to the ultimate craniofacial apostasy—the possibility that operative intervention for sagittal synostosis may not be required at all!³ We addressed the topic tangentially and only because we perceived a tendency to extrapolate arguments promoting “prophylactic” intervention in complex/syndromic synostosis to children with only one affected suture.

Rather than dealing individually with the other points Professor Mathijssen and colleagues raise in their letter (many of which we agree with), we offer the following more general observations.

The prevailing strategy of recommending some form of vault expansion as the default management policy for, in particular, complex/syndromic forms of craniosynostosis (traceable back to the more quoted than actually read 1982 paper by Renier et al.²) can conveniently be expressed as

the hypothesis: “Raised intracranial pressure imposes a sufficiently severe (and predictable) risk upon the brain of the child with craniosynostosis to justify the risks of the surgery and anaesthesia aimed at its relief (or prevention).”

The strength of any hypothesis can be measured by its ability to withstand the attacks made upon it—the stronger the attack it can survive, the stronger the hypothesis. It is our contention that the majority of studies into the connection between raised ICP and NCI have been designed not as attacks upon the hypothesis that underlies it but as ways of propping it up—and therefore weak by definition.

It is instructive to list (in no particular order) the many weakening pitfalls to which such studies often fall victim—sometimes with more than one affecting the same study:

1. Biased selection of patients (exclusions not discussed; studies limited to those who volunteered to be studied)
2. Nonidentical patient cohorts (particularly relevant for “early versus late surgery outcome” studies)
3. Absence of nonoperated controls
4. Variations in age at intervention, in techniques employed, and even in the aims of surgery (shape change vs ICP reduction in single-suture synostosis, for example)
5. Dependence on low sensitivity and/or specificity indicators of ICP as, for example, headache, behavior change, frequent awakenings during the night, and developmental delay—a problem compounded by bundling several together
6. Small numbers observed over short periods or larger numbers accumulated over many years—sometimes by pooling data from multiple centers
7. Lack of independent assessments (i.e., assessments not conducted by those responsible for the intervention under investigation)
8. Variations in both the timing and the methods used for developmental assessments that often cover several age ranges

To summarize, investigations into the connection between ICP and NCI have relied heavily on the “panhandling” method of clinical research: “Dip a sieve into the data and see if it comes up with anything that supports your way of thinking.”

It was because of the prevalence of these and other pitfalls that our paper concluded that for children with craniosynostosis, “The evidence that levels [of ICP] frequently accepted as elevated can be responsible (in the absence of hydrocephalus) for impaired cognitive impairment is weak at best.” It is encouraging therefore to read Professor Mathijssen and her Rotterdam colleagues’ own conclusion that “there is more to craniosynostosis than just ICP.”

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Ventriculoperitoneal shunts after abdominal surgery

TO THE EDITOR: I read with interest the recent article by Burks et al.¹ (Burks JD, Conner AK, Briggs RG, et al: Risk of failure in pediatric ventriculoperitoneal shunts placed after abdominal surgery. *J Neurosurg Pediatr* 19:571–577, May 2017). The authors report that their retrospective review of 141 patients revealed that shunt surgery performed within 2 weeks of abdominal surgery was associated with time to failure, whereas shunt surgery performed 2 weeks after abdominal surgery was not associated with time to failure. They then concluded that when feasible, patients who require shunt placement within the first 2 weeks after an abdominal surgery would probably benefit from either atrial or pleural termini instead.¹ However, I wish to point out that the suggestion of using the pleura in their conclusion would only be applicable to children who are older than 4 years of age.⁴ This is because the absorptive capacity of the pleura is variable and may be insufficient for children who are younger than this age, predisposing them to the development of pleural effusions and respiratory difficulties. For this group of patients, the subgaleal space may probably be considered as a much safer alternative for shunt placement, even though it is commonly used for posthemorrhagic hydrocephalus in premature infants, in addition to the atrium.^{2–4}

Additionally, their study demonstrates an association between shunt placement for hydrocephalus due to spinal dysraphism and shunt failure. This further confirms and strengthens previous findings that suggest that the underlying etiology of hydrocephalus is an important predictor of shunt failure.² The incidence of shunt-related complications following shunting for hydrocephalus due to myelomeningocele alone has been documented to be as high as 22.8%. Unfortunately, most of these risk factors for shunt failure can't be modified. This needs to be taken into consideration during preoperative counseling on the possibility of shunt failure, even with the best of optimal conditions.

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The author reports no conflict of interest.

Response

We appreciate the interest of Dr. Onyia, and are glad for the opportunity to respond. We agree that the pleural space can be less than ideal in infants, but have successfully used pleural shunts in children as young as 2 years, especially as a short-term bridge to entering the peritoneum. The ventriculopleural shunt fell from favor in young children due to its inability to produce favorable long-term results; however, it has been shown to be suitable in the short term.² In our experience, the length of time required after abdominal surgery to bridge for safe placement in the peritoneum is not long enough to overwhelm the pleural absorptive capacity, even in very small children.

Additionally, we have not found subgaleal shunts to be useful for older children except as a very short-term bridge, because therapeutic CSF diversion with a subgaleal shunt typically requires intermittent revisions.⁴

As Dr. Onyia points out, both this work and others have demonstrated a relationship between hydrocephalus etiology and shunt failure.^{1,3} Although efforts to minimize the risk of shunt failure can help drive decisions about shunt placement, we agree that unmodifiable risk factors must indeed also be taken into consideration.

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