Neurosurgical Forum LETTERS TO THE EDITOR

Endoscopic third ventriculostomy with choroid plexus cauterization

TO THE EDITOR: I thoroughly enjoyed reading the article⁴ by Dewan et al. (Dewan MC, Lim J, Morgan CD, et al: Endoscopic third ventriculostomy with choroid plexus cauterization outcome: distinguishing success from failure. *J Neurosurg Pediatr* 18:655–662, December 2016). I congratulate the authors on an extremely well-written, honest, and well-designed study attempting to quantify an outcome that is surprisingly subjective. Surgeons and parents may become emotionally invested in remaining shunt free after such a procedure; placing the shunt may be considered a “defeat.”

While I hope the endoscopic third ventriculostomy/choroid plexus cauterization (ETV/CPC) helps many children, I find it sobering that, even in expert hands, this procedure fails in newborns 60% of the time. The authors report a median age of 0.82 months in the “failure” group, suggesting a high rate of failure in newborns—67% for patients with intraventricular hemorrhage, 66% for those with congenital communicating hydrocephalus, and 78% for those with aqueductal stenosis.

Although placing a shunt certainly has risks, the chances of immediate severe injury or death are exceedingly low. The risk of ETV in experienced hands is also low, although the literature reports a 1% rate of abandoning the case secondary to hemorrhage, a 0.2% risk of fatal basilar artery injury, and an overall complication rate of approximately 8.5%.^{1–3} My personal opinion is that these numbers in the literature are underestimated, underreported, and often do not include any associated morbidity with the CPC.

While I have full confidence in the pediatric neurosurgical community to technically perform this procedure, I think a question that must be addressed is “Should we?” And in whom? Do the authors recommend performing an ETV/CPC in a newborn with aqueductal stenosis, knowing that almost 80% of these operations will fail? All neurosurgeons are painfully aware of the lifelong commitment, morbidity, and potential mortality associated with shunts. However, at some point I would like the leaders of neurosurgery to address the ethics of what failure rate is acceptable to continue to perform this procedure in the very young.

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References

1. Baykan N, Isbir O, Gercek A, Dagnar A, Ozek MM: Ten years of experience with pediatric neuroendoscopic third ventriculostomy: features and perioperative complications of 210 cases. *J Neurosurg Anesthesiol* 17:33–37, 2005
2. Bouras T, Sgouros S: Complications of endoscopic third ventriculostomy. *World Neurosurg* 79 (2 Suppl):S22.e9–S22.e12, 2013
3. Brockmeyer D, Abtin K, Carey L, Walker ML: Endoscopic third ventriculostomy: an outcome analysis. *Pediatr Neurosurg* 28:236–240, 1998
4. Dewan MC, Lim J, Morgan CD, Gannon SR, Shannon CN, Wellons JC III, et al: Endoscopic third ventriculostomy with choroid plexus cauterization outcome: distinguishing success from failure. *J Neurosurg Pediatr* 18:655–662, 2016

Disclosures

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Response

We would like to thank Dr. Fulkerson for his thoughtful and honest remarks on this important topic. In this insightful essay, he notes the discouraging success rate of ETV/CPC in very young infants and asks if what we are doing is right. Should we be subjecting neonates to a procedure that we suspect may make them more likely than not to soon require a subsequent hydrocephalus operation? Are there certain patients who should not be offered ETV/CPC? Before delving into these questions, it is worthwhile to address several features of ETV/CPC introduced in this commentary.

First, the reader should be cautioned not to treat ETV and ETV/CPC as identical procedures with identical outcomes. The literature cited by Dr. Fulkerson is for ETV alone—a different technique indeed. ETV/CPC, which was developed because of the high failure rate of ETV alone among infants, is performed with a flexible endoscope, and electrocautery is never used to penetrate the third ventricle floor. With regard to risk, at the hospital where we both have received training in this technique, there has never been a basilar artery rupture in more than 5000 cases; operative mortality is no greater than for shunt placement. In addition, the infection rate for ETV/CPC is less than that for shunt placement in these infants.

Second, the failure rate in newborns that is described both in our manuscript and in Dr. Fulkerson’s editorial is for short-term failure. It is well established that early failures (< 3 months) are more common for ETV/CPC than for ventriculoperitoneal shunting (VPS). However, unlike VPS, which carries a higher failure rate beyond 3 months,

failure of ETV/CPC after 6 months is rare.¹ Furthermore, failure in the 1st year of life is quite different from failure from the age of 2 years into adulthood. Most ETV/CPC failures occur within a so-called “safe zone” in which failure is a visible diagnosis with a more gradual course, whereas the consequences of a later failure of VPS may be less forgiving.

Hydrocephalus treated with a shunt is a chronic disease. Like any chronic disease, it is susceptible to flares (e.g., shunt malfunctions); it causes patient and caregiver anxiety (e.g., unrelated headaches); and results in tremendous health care expenditures (e.g., radiographic tests, emergency department visits, hospital admissions, and reoperations) over the course of a lifetime. In contrast, a successful ETV/CPC nearly transforms hydrocephalus into a transient condition. Should we not be willing to accept a greater short-term failure rate for ETV/CPC than for VPS?

Nonetheless, as Dr. Fulkerson intimates, at Vanderbilt we have been unwilling to accept a 60% short-term failure rate in neonates—although some may argue that this is ultimately the same risk of reoperation as shunt placement within 2 years. Our patient population was particularly young (median age 1.2 months) relative to similar series, almost certainly contributing to the high failure rate (57%). Other North American series of patients with median ages of 3.2 months⁴ and 2–6 months² report failure rates of 43% and 50%, respectively. In light of these results, we now are resistant to offering ETV/CPC to posthemorrhagic patients and those with congenital hydrocephalus at < 1 month of age, while still performing ETV/CPC at all ages in infants with hydrocephalus associated with spina bifida. By adjusting our patient selection criteria, we expect to see an improvement in the overall success rate.

But is this even the right path to take? Perhaps we should temporize such patients until they reach a certain age, at which point ETV/CPC is offered. What would be the costs and risks associated with this strategy? Are we doing these patients a disservice by denying them the opportunity to live a shunt-free life?

Furthermore, evidence-based patient selection is achievable and essential. We know that patients with a scarred cistern are destined to experience treatment failure, and these patients are now receiving VPS outright.^{3,4} On the other hand, an open cistern improves outcome dramatically, even in the very young. In a North American cohort, the success rate for ETV/CPC in posthemorrhagic hydrocephalus (most cases treated surgically at < 1 month corrected age) with an open cistern has been successful in > 70% at 4-year follow-up; better than for shunts.³ Patients with prepontine scarring who are at high risk for treatment failure can be screened out preoperatively with sagittal T2-weighted fast imaging employing steady-state acquisition (FIESTA) MRI.⁴ As with any operative intervention, proper patient selection is important.

How are we to move forward? The pediatric neurosurgery community would be wise to take pause and heed

the recommendation of Dr. Fulkerson. We must address the ethics surrounding this dilemma in an organized and responsible fashion. Long-term clinical and cognitive outcomes from prospective study design are on the horizon. Although we all eagerly await these results, it seems unlikely that this issue will be put entirely to rest upon their publication. We propose inviting our patients and their families into the clinical science of this very discussion. After all, they carry the burden of shunt and ETV complications, revisions, and comorbidities. What short-term ETV/CPC failure rate are you willing to accept for your child? How important is it to you to avoid shunt dependence?

At the intersection of a caregiver's acceptance of risk and the surgeon's expectation of the benefits and limitations of the treatment resides the acceptable failure rate for ETV/CPC. This dynamic balance is not new. Indeed, it is struck each day when we offer informed consent to our patients and their families before any procedure. Unlike other established interventions, ETV/CPC is still relatively new, so the facts and sentiments surrounding these discussions continue to evolve. As long as the patient remains central in this conversation, we as a community of neurosurgeons will find this balance and our patients will be better off as a result.

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References

1. Kulkarni AV, Drake JM, Kestle JRW, Mallucci CL, Sgouros S, Constantini S, et al: Endoscopic third ventriculostomy vs cerebrospinal fluid shunt in the treatment of hydrocephalus in children: a propensity score-adjusted analysis. *Neurosurgery* 67:588–593, 2010
2. Kulkarni AV, Riva-Cambrin J, Browd SR, Drake JM, Holubkov R, Kestle JRW, et al: Endoscopic third ventriculostomy and choroid plexus cauterization in infants with hydrocephalus: a retrospective Hydrocephalus Clinical Research Network study. *J Neurosurg Pediatr* 14:224–229, 2014
3. Stone SSD, Warf BC: Combined endoscopic third ventriculostomy and choroid plexus cauterization as primary treatment for infant hydrocephalus: a prospective North American series. *J Neurosurg Pediatr* 14:439–446, 2014
4. Warf BC, Campbell JW, Riddle E: Initial experience with combined endoscopic third ventriculostomy and choroid plexus cauterization for post-hemorrhagic hydrocephalus of prematurity: the importance of prepontine cistern status and the predictive value of FIESTA MRI imaging. *Childs Nerv Syst* 27:1063–1071, 2011

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