Chad R. Gordon, DO*‡
Amir Wolff, DMD*‡
Gabriel F. Santiago, MD*‡
Kenneth Liebman, MD§‖
Erol Veznedaroglu, MD§‖
Frank D. Vrionis, MD, MPH, PhD¶
Judy Huang, MD [ORCID]‡
Henry Brem, MD‡
Mark Luciano, MD, PhD‡

*Division of Neuroplastic and Reconstructive Surgery, Department of Plastic and Reconstructive Surgery, Surgery, Department of Plastic and Reconstructive Surgery, Johns Hopkins University School of Medicine, The Johns Hopkins Hospital, Baltimore, Maryland; ‡Department of Neurosurgery, Johns Hopkins University School of Medicine, The Johns Hopkins Hospital, Baltimore, Maryland; §Department of Neurosurgery, Drexel College of Medicine, Philadelphia, Pennsylvania; ¶Marcus Neuroscience Institute, Boca Raton Regional Hospital, Boca Raton, Florida; ‖Global Neurosciences Institute, Department of Neurosurgery, Drexel University College of Medicine, Philadelphia, Pennsylvania

Correspondence:
Chad R. Gordon, DO,
Division of Neuroplastic and Reconstructive Surgery,
Department of Neurosurgery,
Johns Hopkins University School of Medicine,
JHOC, 8th Floor,
601 N. Caroline St.,
Baltimore, MD 21287.
E-mail: cgordon@jhmi.edu

First-in-Human Experience With Integration of a Hydrocephalus Shunt Device Within a Customized Cranial Implant

BACKGROUND: Implantable shunt devices are critical and life saving for hydrocephalus patients. However, these devices are fraught with high complication rates including scalp dehiscence, exposure, and extrusion. In fact, high shunt valve profiles are correlated with increased complications compared to those with lower profiles. As such, we sought a new method for integrating shunt valves for those challenging patients presenting with scalp-related complications.

OBJECTIVE: To safely implant and integrate a hydrocephalus shunt valve device within a customized cranial implant, in an effort to limit its high-profile nature as a main contributor to shunt failure and scalp breakdown, and at the same time, improve patient satisfaction by preventing contour deformity.

METHODS: A 64-yr-old male presented with an extruding hydrocephalus shunt valve and chronic, open scalp wound. The shunt valve was removed and temporary shunt externalization was performed. He received 2 wk of culture-directed antibiotics. Next, a contralateral craniectomy was performed allowing a new shunt valve system to be implanted within a low-profile, customized cranial implant. All efforts were made, at the patient's request, to decrease the high-profile nature of the shunt valve contributing to his most recent complication.

RESULTS: First-in-human implantation was performed without complication. Postoperative shunt identification and programming was uncomplicated. The high-profile nature of the shunt valve was decreased by 87%. At 10 mo, the patient has experienced no complications and is extremely satisfied with his appearance.

CONCLUSION: This first-in-human experience suggests that a high-profile hydrocephalus shunt device may be safely integrated within a customized cranial implant.

KEY WORDS: Hydrocephalus shunt, Cranioplasty, Cranial implant, Shunt complication, Implant extrusion, Scalp wound, Ventriculoperitoneal shunt

Hydrocephalus shunting is one of the most common procedures performed in all types of neurosurgery. More importantly, implanted hydrocephalus shunts, and their accompanying valves, are manufactured in numerous designs/shapes with a common valve thickness between 3 and 10 mm. Most devices are constructed using a combination of rigid

---

ABBREVIATIONS: CAD/CAM, computer-assisted design/manufacturing; **CSF,** cerebrospinal fluid

Supplemental digital content is available for this article at www.operativeneurosurgery-online.com.

---

plastic and/or silicone placed in the subgaleal space external to the skull in close proximity to the ventricle. However, although their material may be modified in composition, all shunt devices are designed for placement within the head at a "nonanatomic" location fraught with complication between the thin, mobile scalp and the strong, immobile skull. Overall, it is the nonanatomical placement of valve hardware, combined with their rigid construction, which may contribute to their high complication rates approaching 40 to 60%. In fact, complication rates are even more staggering (around 70-80%) in those who have undergone at least one previous shunt revision.[1,2]

FIGURE 1. *Preoperative photograph of inaugural patient presenting to neuroplastic surgery clinic with, left-sided scalp wound and extruding shunt valve. One can appreciate the high-profile shunt's visible deformity (represented by dotted black line), as well as the exposed valve (red arrow).*

Shunt-related complications requiring surgical revision are categorized as either "catheter-related" (ie, proximal or distal obstruction) or "scalp-related" (ie, extrusion through scalp, infection, wound dehiscence, etc).[3,4] Despite numerous strategies for reducing infection, it remains the most common complication with an incidence approaching 30%.[5,6] However, each case of "infection" is different, since the true etiology may include (1) bacterial contamination at time of placement, (2) systemic seeding from blood-borne pathogens, or (3) primary scalp dehiscence/shunt extrusion leading to secondary infection. In other words, infection may not be the sole cause of failure, depending on which condition came first. For example, infection may result from secondary contamination following scalp breakdown, and thus, it is sometimes misclassified as "infection" and not as "incisional dehiscence/scalp wound." This sequela occurs from excessive scalp tension over the nonanatomical, high-profile device and long-standing pressure from underneath causing localized ischemia and tissue necrosis.

For instance, the relentless pressure from underneath the scalp leads to tissue necrosis in a manner quite similar to sacral pressure ulcers commonly seen by plastic surgery. Essentially, the necrotic wound begins at the deepest aspect—closest to the source, since its etiology relates directly to pressure and is invisible to the human eye. In the case of hydrocephalus, the rigid structure is the high-profile shunt valve causing an unbalanced pressure from within, similar to the bony prominence causing the constant pressure in the debilitated, bed-ridden patient. As such, the pressure-induced tissue necrosis from the high-profile shunt valve device leads to the unforeseen formation of an open scalp wound (Figure 1). From there, the shunt hardware becomes exposed and bacterial contamination/biofilm formation is unpreventable and equates to eventual hardware removal.[7,8]

Unfortunately, for shunt-dependent patients with severe hydrocephalus, shunt exposure is a major setback. It necessitates

an admission to the neurological intensive care unit with temporary external ventricular drainage. The duration of external ventricular drainage often ranges from 7 to 14 d and includes culture-directed intravenous antibiotics followed by secondary insertion of a new internalized shunt device. As such, for these complex patients who present with shunt valve extrusion/exposure necessitating removal, most would agree that every effort should be made at time of shunt valve replacement to prevent the scalp-related complication from occurring again.

METHODS

A 63-yr-old male presented with a complicated history including obstructive hydrocephalus, cerebellar stroke, syringomyelia, and Arnold-Chiari malformation (treated by decompression and complicated by infected dural patch). His first ventriculoperitoneal shunt was placed 10 yr prior, and since then, had undergone 2 separate revision surgeries for shunt malfunction. Two years later, he presented to us with a chronic, open scalp wound. Further workup revealed that the open scalp wound was related to hardware extrusion (Figure 1). The patient underwent surgery for shunt hardware removal and insertion of temporary externalized ventricular drain. In addition, the scalp wound was excised by neuroplastic surgery and closed with adjacent tissue transfer. The patient remained in the hospital with external ventricular drainage while receiving culture-directed intravenous antibiotics.

During this time, a customized cranial implant (InvisiShunt, Longeviti Neuro Solutions, Hunt Valley, Maryland) was designed specifically to house the exact shunt valve type planned for future implantation by neurosurgery (M.L.). Computer-assisted design/manufacturing (CAD/CAM) was performed in virtual fashion to allow the multidisciplinary team to coordinate exact locations with respect to contralateral shunt placement, planned craniectomy boundaries, and incisional/scalp flap exposure. Of note, the cranial implant was requested to be made out of high-density polyethylene, so that its mechanical properties would allow for intraoperative modification and flexibility as needed to better accommodate the convex curvature of the cranium—as opposed to a more rigid alloplastic material like poly-ether-ether-ketone or poly-methyl-methacrylate. Of note, the upfront cost for this specific implant was institutional-dependent based on prenegotiated terms with the manufacturer. Furthermore, the cost of the implant is a fraction of the cost of a redo shunt and hospitalization.

Following design and fabrication, the customized cranial implant was shipped to our hospital in sterile packing. This process was managed by the hospital. Data collection and review of this case were performed under an active Institutional Review Board protocol. The patient was informed preoperatively of the detailed plan and no additional consent was required for the operation since the hydrocephalus shunt device and customized cranial implant are both FDA approved; the patient consented to publication of their photograph.

At 2 wk, the second-stage surgery was performed on the contralateral side since the patient had communicating hydrocephalus, and in an effort to avoid the previously contaminated field/scarred scalp (**Video**, **Supplemental Digital Content**). The borders of the limited craniectomy were hand-drawn using a marking pen and template, as shown in Figure 2. Of note, the size of the limited craniectomy was oval-shaped and measured approximately 3.5 × 6.5 cm (Figure 3). The size limits of the craniectomy

FIGURE 2. *Intraoperative photograph of customized cranial implant made of high-density polyethylene. In particular, there is an exact fit design incorporated within the customized implant to accommodate the specific shunt valve system being selected by the neurosurgeon.*
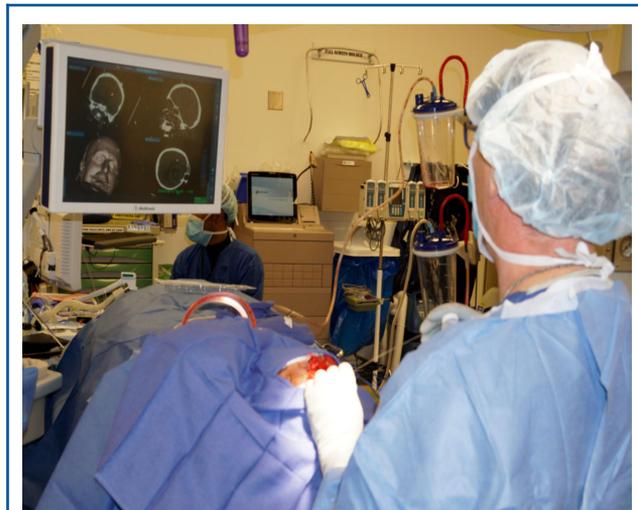

FIGURE 4. *Intraoperative photograph of proximal catheter being placed via navigational guidance.*

FIGURE 5. *Intraoperative photograph of high-profile shunt valve implanted within customized cranial implant along right-sided temporoparietal location. Titanium screws fixate the cranial implant along the periphery.*
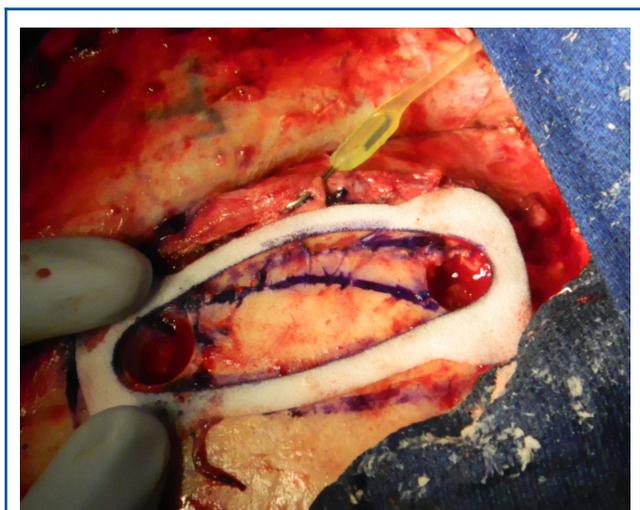

FIGURE 3. *Intraoperative photograph of marking template being used to identify exact burr hole locations on either end. Next, a craniotome is used to complete the limited craniectomy.*

were determined based on length and width of the device. The proximal catheter was placed uneventfully in the lateral ventricle using intraoperative guidance (Figure 4). The distal catheter was placed in the peritoneal cavity using standard technique. Once both catheters were in proper position, the customized cranial implant was inset within the limited craniectomy defect and fixated with titanium screws along the periphery (Figure 5). The high-profile shunt device was then integrated within the implant, and the proximal catheter was passed through and connected. The distal catheter was connected and positioned within a customized groove at an angle less than 90 degrees. Following integration, the shunt valve was measured before inset to be 8 mm tall. Following implantation within the customized cranial implant, its relative height was reduced to just 1 mm, which left its edges just high enough to palpate

FIGURE 6. *Intraoperative photograph showing the re-draping and inset of a pericranial-temporalis muscle flap over the inferior shunt valve/cranial implant in preparation for final scalp closure.*

FIGURE 7. *Right-sided intraoperative photograph of the final cranial contour following multilayered scalp closure. Of note, a single suture (red arrow) was placed above the low-profile shunt valve for this inaugural case to assist with postoperative device identification.*

subcutaneously with one's fingers. Proper function and shunt flow were confirmed prior to flap inset, multilayered scalp closure, and closed suction drain insertion (Figures 6 and 7).

RESULTS

The patient underwent an uneventful recovery. At 9 mo, the patient has experienced no scalp wound complications and/or pain in the area of the device. In particular, the surrounding
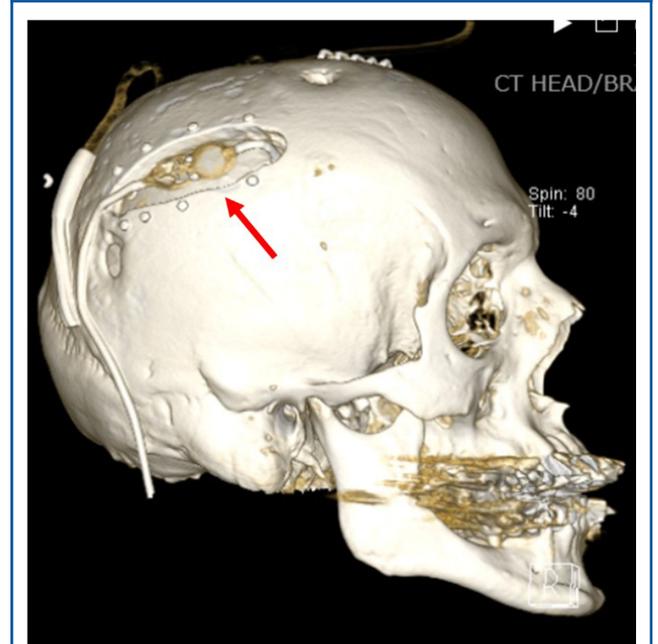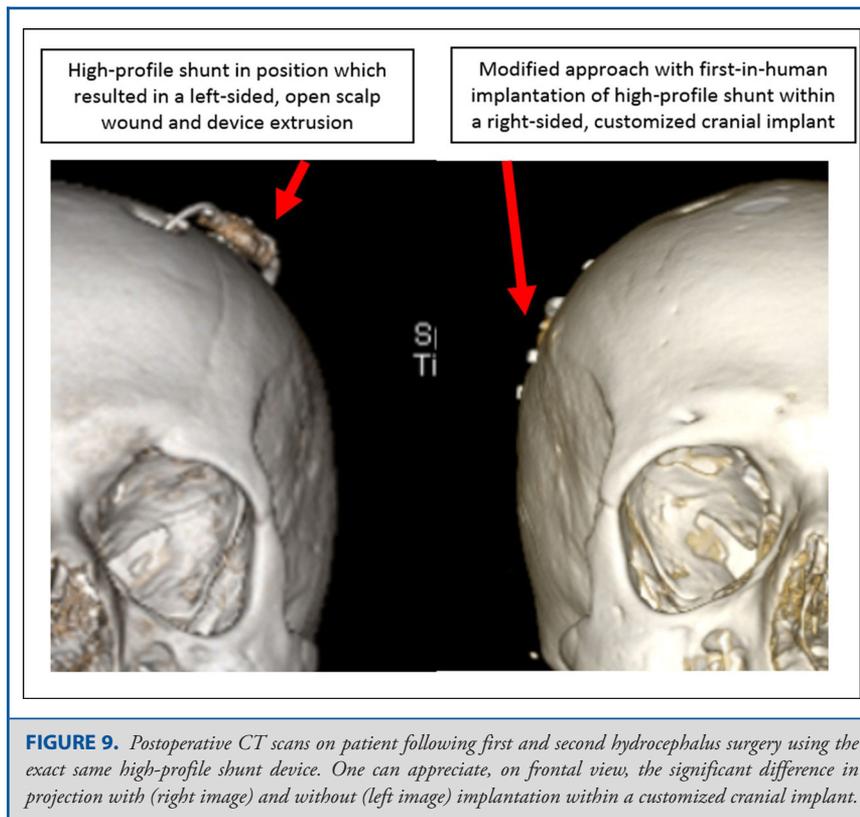

FIGURE 8. *Postoperative CT scan with 3-dimensional reconstruction showing right-sided hydrocephalus shunt valve implantation within a customized cranial implant. In particular, the high-density polyethylene cranial implant is radiolucent and cannot be seen within the craniectomy site.*

cranial implant has not interfered with any shunt programming or valve localization. To date, his hydrocephalus has remained well controlled. In particular, the overall height of his shunt valve device was decreased by 87%, as compared to its previous placement height which led to eventual extrusion (Figure 8). Overall, there is a significant difference in shunt valve prominence as compared to its previous placement, as shown in Figure 9.

DISCUSSION

Although shunt failure has been extensively studied, most studies have focused on pediatric patients.[9] With this in mind, we present our experience with shunt-related scalp complications in the adult population. Furthermore, we present a new approach to lower shunt device profiles, especially in those who present with hardware extrusion and/or incisional scalp dehiscence. One well-described option is to shave down some cortical bone in a shape that matches the device. However, this approach is time consuming and only reduces the height by a maximum of 1 to 2 mm.[10] As such, we chose to instead perform a limited, full-thickness craniectomy in an effort to drastically lower the shunt's profile and to minimize risk for repeat extrusion. Of note, this technique has been performed successfully in other similar settings where implants are of high profile, such as deep brain stimulators, plates, and cortical stimulators.

FIGURE 9. *Postoperative CT scans on patient following first and second hydrocephalus surgery using the exact same high-profile shunt device. One can appreciate, on frontal view, the significant difference in projection with (right image) and without (left image) implantation within a customized cranial implant.*

Approximately 40 000 new shunts are placed annually for hydrocephalus management. Unfortunately, shunt replacement and shunt removal are quite frequent at a rate of 43% and 7%, respectively.[5] In fact, Reddy et al[9] investigated predisposing risk factors for all shunt-related complications in 1015 patients and found the rate of revision shunt surgery to be close to 1 out of every 2 patients (46%). Furthermore, the history of previous shunt revision surgery increases the odds of needing subsequent surgery by 9-fold, and when revision surgery is required, it occurs most often within the first 6 mo. Interestingly, they found that "infection-related complications" are the most common cause for re-admission in shunted hydrocephalus patients.[9] However, overall shunt infections are more likely secondary to initial intention rather than scalp breakdown, and so further investigation is needed to estimate the incidence of both scenarios.

Given the staggering rate (46-48%) for shunt revision in one's lifetime, novel or unconventional strategies by which these complications could be minimized and/or avoided altogether are worthy of consideration.[4,11] In looking at the current design of shunts, our team noted a common design characteristic being the high-profile nature of the shunt valve, which is placed between the rigid skull and supple scalp. In our opinion, devastating complications such as scalp wounds and implant exposure/infection are foreseeable when using high-profile, extra-anatomical shunt valves, simply because these devices place unsafe pressure on the deep scalp, where its vascular perfusion is most affected (ie, dermal-subdermal plexus). This hypothesis is confirmed by other studies that show that high-profile shunts, as opposed to low-profile shunts, have a greater tendency to require removal secondary to extrusion/infection.[7,8]

With regard to shunt re-implantation for those patients with history of scalp-related complications, our preference is to place all hardware (shunt tubing and valve) opposite the side of the original infection, if possible, in cases of communicating hydrocephalus. Either way, we strive to relocate all replaced hardware in an area most distal from the previous compromised scalp. Thus, when bilateral ventricular dilation or communicating hydrocephalus is noted, we use the contralateral side. This strategy serves 2 benefits. First, it helps avoid the previously contaminated soft tissue pocket. Second, it removes the permanent shunt from the zone of injury (ie, scar tissue), thereby allowing us to place the new hardware under healthy scalp with normal perfusion and thickness.

In this particular instance, the main precipitating factor was the high-profile, rigid shunt device and its pressure on the overlying scalp. Therefore, to address this, we performed a small-sized craniectomy using a burr and craniotome (by way of a predesigned marking template supplied by the manufacturer). Attempting to drill a partial-thickness cranial defect to mirror the necessary shunt device would have been quite challenging given

its 3-dimensional shape. Furthermore, a partial-thickness defect may have only reduced the overall thickness by 1 to 2 mm.[12] As such, following a prolonged discussion with the patient in regard to the novelty of this approach, we decided to use a low-profile customized implant to reduce the valve's height by 87%, and at the same time, eliminate all concern for shunt migration by providing a "key-and-lock" type fit (Figure 5).

While the hydrocephalus shunt is credited with saving and improving millions of lives worldwide each year, there are also challenging associated complications. Therefore, examination of shunt systems in order to help minimize co-existing variables, which lead to extraordinarily high revision rates, may be warranted. The proposed method of reducing shunt valve prominence avoids compression of the scalp's vital blood supply. In addition, the use of a customized cranial implant is attractive as a "manufacturer-agnostic" alternative, meaning that the CAD/CAM process can shape it for all shunt valve types.

Regardless, we predict that design changes such as these may serve to significantly decrease the need for shunt-related revision surgery and the cumulative effects of shunt revision surgery/interval hospital admissions. It is estimated that the average cost of a cerebrospinal fluid (CSF) catheter-related infection in the United States is approximately $50 000. Therefore, it is imperative to consider these consequences when assessing the upfront cost of customized implant materials designed to prevent revision surgery.[6, 13] Another advantage (well appreciated by this patient) in reducing valve prominence is the minimization of visible deformity along the scalp when compared with its usual placement (Figure 10). Of note, this is increasingly important in patients with male-pattern balding and/or short hair, who hope to avoid the social stigma accompanying their neurological condition and/or the fact that they have undergone previous brain surgery.

Unquestionably, shunt-related complications are devastating to all patients regardless of age, ranging from simple adjustment to definitive removal. Although these complications are complex and multifactorial, the scalp-related variables play a major factor in cases of extrusion/failure and therefore, with this advance, could be prevented by way of a low-profile inset. As such, the ability to securely house shunt valves within the confines of a craniectomy defect (ie, intercranial space) by taking advantage of underutilized space within a cranial implant provides many newfound advantages (Figure 11).[14-16] This includes offering our patients an option for an improved appearance with enhanced contour, while concurrently decreasing detrimental pressure and relative ischemia onto their overlying scalp (Figures 8 and 9).

Limitations

Of note, there are several limitations to this preliminary report. First, it is limited to just a single patient. However, at the time of this submission, a total of 8 cases have been performed with no complications to date as part of a larger multicenter study
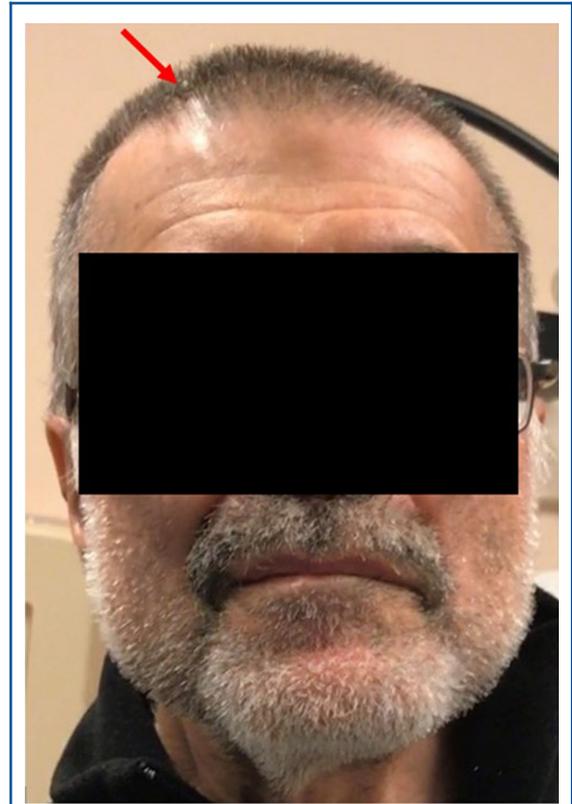

FIGURE 10. *Frontal view of inaugural patient at 1 mo following right-sided hydrocephalus shunt valve implantation within a customized cranial implant. In addition, there is no visible contour deformity other than a small palpable bump.*

involving various neurosurgical groups. Second, a relatively short follow-up time of 9 mo may not capture eventual complications which could develop at a later date, and thus a large series of successful implantations is unquestionably warranted. However, given the variables which we modified to prevent future "scalp-related" complications, in combination with data showing that shunt-related complications most often occur within the first 6 mo, we feel obliged to share our initial experience with others challenged by shunt valve extrusions/complicated scalp wounds and the limited options which currently exist.[4]

Advantages and Disadvantages

In parallel, there are several important advantages and disadvantages with this first-in-human experience. First, the advantages of integrating hardware within a customized cranial implant is an advance and may serve as a platform for all hydrocephalus-related neurotechnologies, including flow meters to detect shunt malfunction and/or pressure monitors, to diagnose elevated intracranial pressures. Of course, as this research progresses, one could envision implanting some of these technologies all
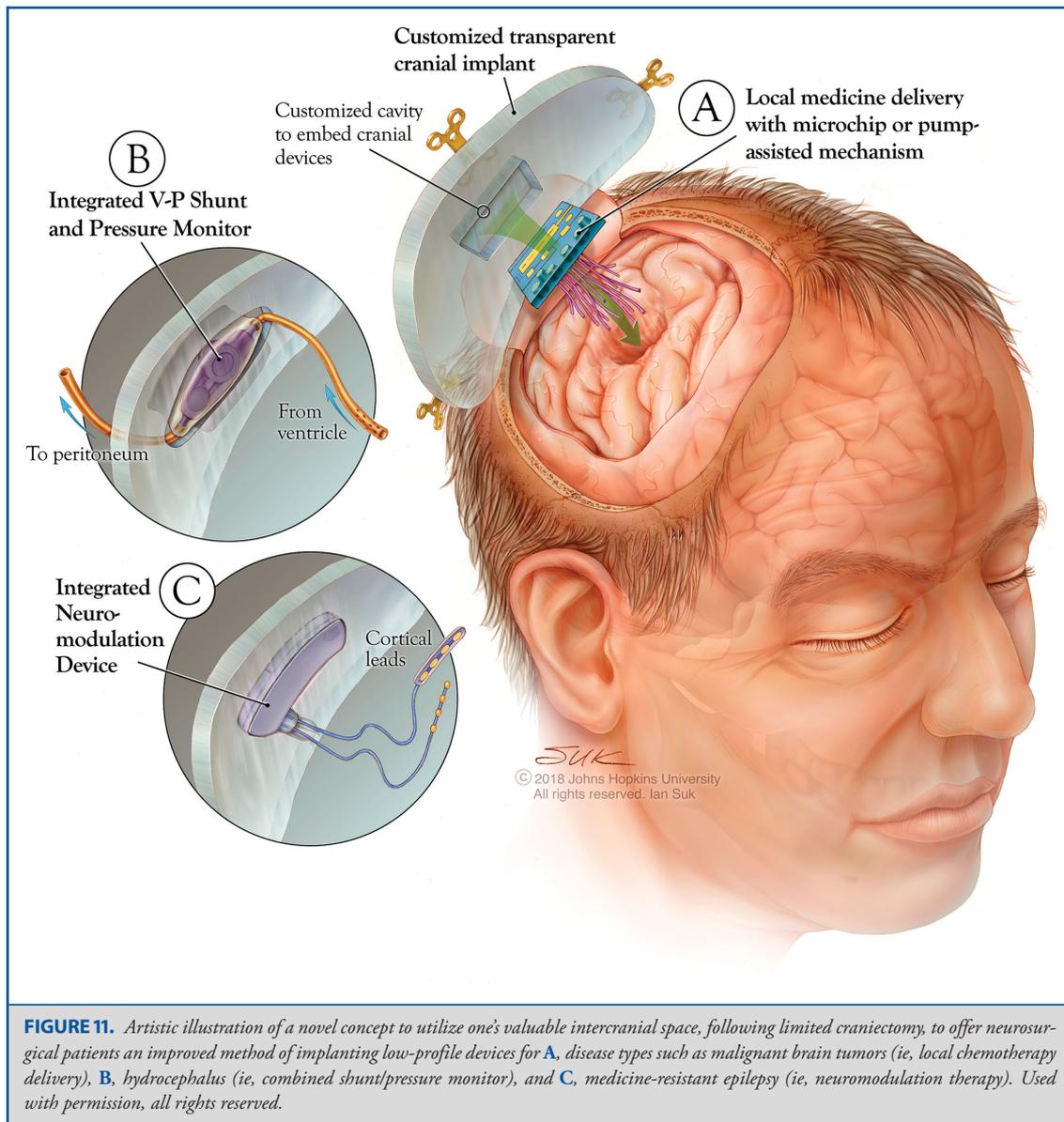
FIGURE 11. *Artistic illustration of a novel concept to utilize one's valuable intercranial space, following limited craniectomy, to offer neurosurgical patients an improved method of implanting low-profile devices for* **A**, *disease types such as malignant brain tumors (ie, local chemotherapy delivery),* **B**, *hydrocephalus (ie, combined shunt/pressure monitor), and* **C**, *medicine-resistant epilepsy (ie, neuromodulation therapy). Used with permission, all rights reserved.*

at the same time. In contrast, the obvious disadvantage is the craniectomy which is required, as opposed to a standard burr hole for proximal catheter insertion, which is greater in dimension. Thus, there is additional risk for related complications such as durotomy, CSF leak, and epidural hematoma. However, in an instance such as the one presented here—in a patient with previous shunt hardware extrusion—the risk-to-benefit ratio of providing a low-profile cranial implant to prevent further scalp-related complications vs the accompanying craniectomy risks is well justified. As a result, this approach should be considered most applicable for cases with recurrent shunt failure and with vulnerable scalp.

CONCLUSION

The prominence of hydrocephalus shunt devices under the scalp by way of standard implantation is a contributing factor for soft tissue complications associated with hydrocephalus. Integration of the high-profile shunt valve within the confines of a customized cranial implant appears to be both safe and reliable. This first-in-human experience provides a pragmatic strategy for (1) minimizing risk (ie, open scalp wounds/extrusion), (2) providing a novel strategy to improve long-term outcomes, (3) increasing the potential to optimize patient satisfaction and appearance, and (4) supplying a viable platform for exciting future

neurotechnology advances related to hydrocephalus such as flow meters and pressure monitors.

Disclosures

Dr Gordon is a consultant for Stryker and Longeviti Neuro Solutions. Dr Huang and Gordon are stockholders in Longeviti. The other authors have no personal, financial, or institutional interest in any of the drugs, materials, or devices described in this article.

REFERENCES

1. Kofoed Mansson P, Johansson S, Ziebell M, Juhler M. Forty years of shunt surgery at Rigshospitalet, Denmark: a retrospective study comparing past and present rates and causes of revision and infection. *BMJ Open*. 2017;7(1): e013389.
2. Reddy KG, Papireddy Bollam P, Caldito G. Ventriculoperitoneal shunt surgery and the risk of shunt infection in patients with hydrocephalus: long-Term single institution experience. *World Neurosurg*. 2012;78(1-2):155-163.
3. Kast J, Duong D, Nowzari F, Chadduck WM, Schiff SJ. Time-related patterns of ventricular shunt failure. *Child's Nerv Syst*. 1994;10(8):524-528.
4. Reddy GK, Bollam P, Caldito G. Long-term outcomes of ventriculoperitoneal shunt surgery in patients with hydrocephalus. *World Neurosurg*. 2014;81(2): 404-410.
5. Zabramski JM, Whiting D, Darouiche RO, et al. Efficacy of antimicrobial-impregnated external ventricular drain catheters: a prospective, randomized, controlled trial. *J Neurosurg*. 2003;98(4):725-730.
6. Edwards NC, Engelhart L, Casamento EM, McGirt MJ. Cost-consequence analysis of antibiotic-impregnated shunts and external ventricular drains in hydrocephalus. *J Neurosurg*. 2015;122(1):139-147.
7. Nguyen TA, Cohen PR. Scalp necrosis overlying a ventriculoperitoneal shunt: a case report and literature review. *Dermatol Online J*. 2015;21(10):13030/qt2rs544f9.
8. Joo JD, Jang JU, Kim H, Yoon ES, Kang DH. Cranial defect overlying a ventriculoperitoneal shunt: pressure gradient leading to free flap deterioration? *Arch Craniofac Surg*. 2017;18(3):186-190.
9. Khan F, Rehman A, Shamim M, Muhammad B. Factors affecting ventriculoperitoneal shunt survival in adult patients. *Surg Neurol Int*. 2015;6 (1):25.
10. Cakir M. "Reverse J Shaped" skin flap and cranial bone groove for shunt hardware: a new technique in patients with fragile skin. *Turk Neurosurg*. published online ahead of print: July 11, 2017 (doi: 10.5137/1019-5149.JTN.20444-17.1).
11. Wang JY, Jackson EM, Jallo GI, Ahn ES. Shunt revision requirements after posthemorrhagic hydrocephalus of prematurity: insight into the time course of shunt dependency. *Childs Nerv Syst*. 2015;31(11):2123-2130.
12. Ammar A, Nasser M. A long-term complication of burying a shunt valve in the skull. *Neurosurg Rev*. 1995;18(1):65-67.
13. Asemota A, Santiago GF, Zhong S, Gordon CR. Comparative cost analysis of single and mutli-stage temporal deformity correction following neurosurgical procedures. *J Craniofac Surg*. 2018;29(1):130-138.
14. Gordon CR, Santiago GF, Huang J, Bergey GK, Liu S, Armand M, Brem H, Anderson WS. First in-human experience with complete integration of neuromodulation device within a customized cranial implant. *Oper Neurosurg*. 2018;15(1):39-45.
15. Gordon C. Low-profile intercranial device. *US Patent Application*. Published September 3, 2017. Assignee: The Johns Hopkins University.
16. Gordon C, Christopher J. Method for manufacturing a low-profile intercranial device and the low-profile intercranial device manufactured thereby. *US Patent Application*. Published March 1, 2018. Assignee: The Johns Hopkins University.

Supplemental digital content is available for this article at www.operativeneurosurgery-online.com.

Supplemental Digital Content. Video. Summary video demonstrating surgical details and placement of customized cranial implant for integration of hydrocephalus shunt device.