The term “syndrome of the trephined” was coined in the 1930s to describe a variety of symptoms in patients with cranial defects that were relieved by cranioplasty “in a large majority of cases.”4 The term “syndrome of the sinking skin flap” was applied to a subset of patients with large concave cranial defects after decompressive craniectomy who experienced spontaneous neurological deterioration, some of whom showed improvement after cranioplasty.22 In more recent series, both retrospective and prospective, improvement in neurological function after cranioplasty was reported in 16%–26%.7,18 We cared for 2 patients with postrumatic decompressive craniectomy whose marked deterioration caused by so-called paradoxical herniation16 was not spontaneous but provoked, one by a lumbar puncture and the other by a ventriculoperitoneal shunt. These 2 patients prompted us to review our experience and those in the literature.

Methods

We describe here both index cases, to whom we refer as Patients A and B to ensure anonymity. We also investigated a series of consecutive patients at a single hospital who underwent decompressive craniectomy for traumatic brain injury over a specific time interval to identify those who also underwent a lumbar puncture or ventriculoperitoneal shunting.
bar puncture or ventriculoperitoneal shunting between the
time of the decompressive craniectomy and cranioplasty,
death, or loss to follow-up. We documented those whose
ccondition deteriorated after the procedure to gain some
understanding of the frequency of this complication and
of the factors related to the deterioration. Finally, we re-
viewed the literature for similar cases by searching the
phrases quoted in the introduction in PubMed and the bib-
liographies of all papers so identified.

Results

Patient A: Index Case of Deterioration After Lumbar
Puncture

This patient was struck by a car while riding his bicycle
without a helmet. He sustained skull fractures, a right-sid-
ed subdural hematoma, and a right frontal contusion for
which a right-sided decompressive craniectomy was per-
formed soon after admission. The patient was discharged
to a rehabilitation facility approximately 1 month after
the accident. When readmitted for a cranioplasty approxi-
mately 3 months after the accident, the patient was noted
to be alert and oriented, and he had residual weakness and
pain on the left side. The skin flap was sunken on the right.
The planned procedure was cancelled because of a uri-
nary tract infection. A cranioplasty was performed at a
later admission, but the bone became infected and had to
be removed. The patient continued to improve neurologi-
cally and participated in inpatient rehabilitation. Apprrox-
imately 1 year after the initial injury, the patient returned
to the hospital with a fever and elevated white blood cell
count. A lumbar puncture was performed as part of the
fever evaluation; meningitis was not found. The patient’s
condition subsequently deteriorated, and he became com-
motose over the ensuing 24 hours. Serial imaging revealed
paradoxical herniation (Fig. 1). Despite treatment with flu-
ids, positioning with his head down, and a blood patch,
the patient did not improve neurologically to the level of
functioning before the lumbar puncture. When last seen
approximately 2 months after the lumbar puncture, the
patient was minimally responsive but able to follow some
simple commands on the right side.

Patient B: Index Case of Deterioration After
Ventriculoperitoneal Shunting

This patient fell from a skateboard without a helmet
and sustained skull fractures, a left-sided subdural hema-
toma, and bilateral frontal contusions for which a left-sid-
ed decompressive craniectomy was performed soon after
admission. Initially, the patient was recovering well but
then worsened after approximately 2 weeks. The patient
was found to have hydrocephalus, and a right frontal ven-
triculoperitoneal shunt with an adjustable pressure valve
was placed. After intensive inpatient rehabilitation, the
patient was discharged home approximately 6 weeks af-
fter the injury. Between 2 and 3 months after the accident,
the patient began to experience increasing right-sided
weakness and word-finding problems. The skin flap was
sunken on the left. These problems then worsened, and
imaging revealed paradoxical herniation (Fig. 2). The pa-
tient’s weakness and aphasia improved after adjustment of
the shunt pressure valve to reduce drainage and eventually
disappeared after a cranioplasty, which was performed ap-
proximately 5 months after the accident.

Case Series at Harborview Medical Center

After securing approval from the University of Wash-
ington Human Subjects Committee, we used adminis-
trative databases to identify each patient who underwent
decompressive craniectomy for a traumatic brain injury
at Harborview Medical Center in Seattle, Washington,
before November 1, 2006. As part of the procedure, the
dura is always opened with a stellate-type incision to per-
mit brain herniation through the craniectomy defect. The
medical records of 205 patients were screened to find doc-
umentation that spinal fluid was assayed or that a ventricu-
loperitoneal shunt was placed after decompressive crani-
etomy and before cranioplasty. Details about the lumbar
puncture were often lacking. All analyses were conducted
using Stata version 13.1 (StataCorp).

The initial case in the series was from July 15, 1991.
The index case for the lumbar puncture was included
among the group of 205 patients, whereas the index case
for shunting was not included because treatment occurred
after 2006. The screen yielded 36 patients who underwent
lumbar puncture and 10 patients who underwent ventricu-
loperitoneal shunting who met these criteria. Deteriora-
tion that resulted from paradoxical herniation occurred in
2 of the 36 patients after lumbar puncture (5.6% [exact
binomial 95% CI 0.7%–18.7%]) and in 3 of the 10 patients
after ventriculoperitoneal shunting (30% [exact binomial 95% CI 6.7%–65.2%]).

The number of days between the decompressive craniectomy and the last lumbar puncture, for those who underwent >1, was significantly higher for those who deteriorated than for those who did not (mean 206 vs 31 days, respectively; p < 0.05 by t-test and the Wilcoxon rank-sum test). Most early lumbar punctures were performed to either control intracranial pressure or evaluate fever, whereas most late lumbar punctures were performed to evaluate fever. Only 10 patients had lumbar puncture >30 days after the decompressive craniectomy, and 2 of them deteriorated (20% [exact binomial 95% CI 2.4%–55.6%]). Both of these patients deteriorated within 24 hours of the lumbar puncture.

Brain CT images obtained within 48 hours of the lumbar puncture were available for 9 of these 10 patients; the remaining patient’s CT images were obtained 5 days after the lumbar puncture. These CT scans were reviewed for midline shift, the size of the lateral and third ventricles, and the appearance of the skin flap, which has been characterized as sunken, flat, full, and bulging (see Table 1 in the paper by Yamaura and Makino). We compared these variables between the 2 patients who deteriorated after the lumbar puncture and the 7 patients who did not. Only 2 variables were significantly different: midline shift and the appearance of the skin flap. The mean midline shift was 0 mm in the 2 patients who deteriorated and 5.4 mm in those who did not deteriorate (2-tailed p < 0.05 by t-test and the Wilcoxon rank-sum test). The shift was toward the side of the decompressive craniectomy except in 1 patient who had a large extraaxial fluid collection on the side of the craniectomy. The skin flap was flat or full before the lumbar puncture in the 2 patients who deteriorated and bulging (n = 6) or full (n = 1) in those who did not (p < 0.5 by the chi-square test). None had a sunken appearance.

With respect to shunts, all 10 patients had them placed contralateral to the side of the decompressive craniectomy. The number of days between the decompressive craniectomy and the ventriculoperitoneal shunting and between the shunting and the cranioplasty or death did not differ significantly between those who did and did not deteriorate. In the 3 patients who deteriorated, changes in imaging were variable and occurred as early as 7 days and as late as 62 days after placement of the shunt. At the time the shunt was placed, the craniectomy site was convex with associated hydrocephalus. By the time of deterioration, the site was concave, with the sunken skin flap and paradoxical herniation evident on imaging.

**Literature Review**

In 1969, Yamaura and Makino first observed the syndrome of the sinking skin flap in a patient who spontaneously deteriorated, which prompted them to study 33 patients who underwent decompressive craniectomy for a variety of reasons. To understand better the improvement seen after cranioplasty, they described lumbar punctures performed in a subset of 13 patients immediately before cranioplasty. With a single exception, the opening pressure was low. No deterioration after the lumbar puncture was observed, but cranioplasty was performed soon thereafter and resulted in improvement in 4 of these 13 patients (31%). In a subsequent study, 40 patients with craniectomy underwent lumbar puncture with hydrodynamic studies both before and after cranioplasty. Of these patients, 36 had symptoms, and in 22 of these patients the symptoms were believed to be related to the syndrome of the trephined. The lumbar punctures were performed, on average, 6.5 days before the cranioplasty. The opening pressure was found to be low in all the patients with symptoms and to increase significantly after cranioplasty, but none of the patients was described as deteriorating after the lumbar puncture, which entailed the infusion of artificial cerebrospinal fluid.

In what is likely the first report of provoked paradoxical herniation, deterioration in 2 patients after craniectomy for tumor resection was described and believed to be secondary to an ongoing leak from the lumbar puncture site. The lumbar punctures had been performed to facilitate tumor resection by lowering elevated intracranial pressure. Deterioration was reversed in these patients with a blood patch. The authors proposed that exposure of the brain to atmospheric pressure allowed for the paradoxical herniation (see Fig. 1 in the paper by Guido and Patterson). Subsequently, 13 more patients with paradoxical herniation provoked by lumbar puncture were described in a series of 4 cases, whose authors first used the term “paradoxical herniation,” and in 9 single case reports. Cases of paradoxical herniation provoked by lumbar puncture are summarized in Table 1. Although the timing was not always easy to determine, lumbar puncture was typically performed >1 month after the craniectomy, which had been performed for a variety of conditions.
including encephalitis, abscess, tumor, ischemic stroke, and trauma. The earliest lumbar puncture in a patient with ischemic stroke or trauma was performed 14 days after decompressive craniectomy in a patient who did not experience severe brain swelling and whose intracranial pressures were normal. The lumbar puncture at 14 days was performed to address a large subgaleal fluid collection and ventricular enlargement. With one exception, the deterioration provoked by the lumbar puncture was evident within minutes to 5 days. In the exceptional case, a lumbar puncture was performed 5 days after decompressive craniectomy for brain swelling related to a subarachnoid hemorrhage. Paradoxical herniation did not occur until 38 days after the lumbar puncture; the cause was presumed to be a lumbar leak.

Most of the cases showed evidence for ventricular enlargement at the time of the lumbar puncture. A recent patient treated with a lumbar puncture followed by a lumbar drain had a bulging skin flap that sank after the drainage of spinal fluid.

Cases of paradoxical herniation provoked by ventriculoperitoneal shunting are summarized in Table 2. In their series of 33 patients, Yamaura and Makino also described a patient who underwent decompressive craniectomy as part of meningioma resection (Case 18). She developed hydrocephalus and required a shunt 6 weeks after the initial surgery. She subsequently deteriorated slowly but showed marked improvement after cranioplasty performed approximately 1 year after her initial surgery. In another series reported by Schiffer et al., a patient deteriorated more immediately after receiving a ventriculoperitoneal shunt and improved after cranioplasty (Case 4). Oh et al. described deterioration after ventriculoperitoneal shunting in 2 of 13 patients with hydrocephalus who were convex at the craniotomy site before the procedure. “In 2 cases with slit ventricle, one week later, the craniectomy site became excessively sunken, followed by neurological deterioration and aggravated concavity of the scalp flap, especially in an upright position. They improved significantly after changing shunt device (programmable shunt device) and cranioplasty.” Two other cases with similar courses were described. A likely seventh case was described in a review of decompressive craniectomy, although the details were scant (see Fig. 6 in the paper by Akins and Guppy).

### Discussion

Although spontaneous neurological deterioration can occur in patients with decompressive craniectomy, both lumbar puncture and ventriculoperitoneal shunting can provoke deterioration that results from paradoxical herniation. The results of our study of a small series of patients who underwent decompressive craniectomy prompted by trauma suggest that the risk of provoked paradoxical herniation after lumbar puncture may be substantial (20% [95% CI 2.4%–55.6%]) if > 1 month has passed since the initial injury. Timing may be less important in patients who undergo placement of a ventriculoperitoneal shunt, because the onset of deterioration may be variable.
TABLE 2. Summary of the literature on patients with deterioration after decompressive craniectomy provoked by ventriculoperitoneal shunting

<table>
<thead>
<tr>
<th>Authors &amp; Year (no. of cases)</th>
<th>Disease Prompting Decompression</th>
<th>Enlarged Ventricles</th>
<th>Days to Shunting</th>
<th>Comments</th>
</tr>
</thead>
<tbody>
<tr>
<td>Yamaura &amp; Makino, 2008</td>
<td>Meningioma removed</td>
<td>Yes</td>
<td>42</td>
<td>Case 18 in the series</td>
</tr>
<tr>
<td>Schiffer et al., 1997</td>
<td>Meningioma removed</td>
<td>Yes</td>
<td>Unknown</td>
<td>Case 4 in the series</td>
</tr>
<tr>
<td>Oh et al., 2008 (2)</td>
<td>Acute injury</td>
<td>Yes</td>
<td>Unknown</td>
<td></td>
</tr>
<tr>
<td>Han et al., 2008</td>
<td>Trauma</td>
<td>Yes</td>
<td>28</td>
<td></td>
</tr>
<tr>
<td>Akins &amp; Guppy, 2008</td>
<td>Unknown</td>
<td>Yes</td>
<td>Unknown</td>
<td>Case described in Fig. 6 of the report</td>
</tr>
<tr>
<td>Wee &amp; Kuo, 2014</td>
<td>Trauma</td>
<td>Yes</td>
<td>~30</td>
<td></td>
</tr>
<tr>
<td>Current series</td>
<td>Trauma</td>
<td>Yes</td>
<td>32</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Trauma</td>
<td>Yes</td>
<td>42</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Trauma</td>
<td>Yes</td>
<td>124</td>
<td></td>
</tr>
<tr>
<td>Index case</td>
<td>Trauma</td>
<td>Yes</td>
<td>14</td>
<td>See Patient B: Index Case of Deterioration After Ventriculoperitoneal Shunting</td>
</tr>
</tbody>
</table>

depending on how much fluid is drained. Again, the risk in our small series of patients after shunt placement was substantial (30% [95% CI 6.7%–65.2%]). In their seminal work, Yamaura and Makino described the skin flap in patients after decompressive craniectomy as sunken, flat, full, or bulging. Although the skin flap may be sunken in spontaneous paradoxical herniation, it is typically flat or full, but not bulging, in provoked paradoxical herniation before lumbar puncture. In fact, the patients in most provoked-herniation cases (Table 1 and Fig. 1) showed evidence of ventricular enlargement before lumbar puncture and thus lacked a sunken skin flap. In our series, 7 patients with a bulging skin flap tolerated a lumbar puncture without deterioration, but based on a recent case report, such an appearance of the skin flap does not afford absolute protection.

The longer the interval between craniectomy and cranioplasty, the greater the risk for spontaneous deterioration, and the more opportunities potentially exist to provoke deterioration. The increased risk over time may exist especially for patients with trauma. Soon after a trauma that requires decompressive craniectomy, the brain is swollen and the skin flap bulges. In our series, 26 of the 36 patients had undergone lumbar puncture at this stage, and none showed evidence of paradoxical herniation. Conversely, brain swelling decreases over time, and 2 of the remaining 10 patients who had undergone lumbar puncture after 1 month had paradoxical herniation; both of these patients had no midline shift and lacked a bulging skin flap on CT images acquired before the lumbar puncture. In situations other than trauma, the risk may be more immediate, as in the first described patients whose deterioration was believed to be secondary to a spinal fluid leak from a lumbar puncture with a lumbar drain used during brain tumor resection.

Nakamura et al. may have been the first to suspect a potential risk from ventriculoperitoneal shunting in patients with craniectomy. They described a patient who had a large decompressive craniectomy after trauma. The patient required a ventriculoperitoneal shunt for hydrocephalus and eventually had a cranioplasty. The implanted plate became infected and had to be removed. The authors noted an immediate neurological deterioration when the patient was upright that could be reversed with the patient lying flat or in the Trendelenburg position. They commented, “The presence of a well-working shunt system is thought to be one of the most important factors in the mechanism which was involved in the development of such marked concave deformity in an erect position.” Deterioration may occur sooner or later after placement of a ventriculoperitoneal shunt depending on the dynamic between the regression of brain swelling and the amount of fluid drained. For example, in our index case, although the shunt was placed early after the trauma and decompressive craniectomy, deterioration was delayed, presumably reflecting the slow regression of brain swelling. Improvement followed reprogramming of the shunt to reduce drainage and a cranioplasty.

The mechanism of paradoxical herniation likely entails exposure of the brain to atmospheric pressure. Low rather than high intracranial pressures ensue, which result in sinking of the skin flap and paradoxical herniation. Because the herniation relates to low pressures, treatments aimed at lowering intracranial pressures may worsen the situation. Various treatments for acute deterioration, including intravenous fluids, positioning with the head down, clamping devices that drain cerebrospinal fluid, infusing fluid into the intrathecal space, and performing a blood patch, have been suggested but are unproven. Regardless of whether deterioration is spontaneous or provoked, improvement can be seen after cranioplasty.

Our study was limited by examining retrospectively at a single institution a single indication for decompressive craniectomy, namely trauma. As a consequence, the numbers are small, and the generalizability is compromised. Nonetheless, most previous investigations have been limited to case reports or small case series. The deterioration provoked by ventriculoperitoneal shunting has not been reported as frequently or with as much detail as that provoked by lumbar puncture. This relative underreporting may be a result of the delay in deterioration that may follow shunting as opposed to the lumbar puncture, as il-
Illustrated by our 2 index cases. More studies are needed to clarify these and other issues related to provoked paradoxical herniation. The ideal study would be a large, prospective, multicenter investigation to examine multiple indications for decompressive craniectomy, which is a study that would pose substantial challenges.

Conclusions

The risk of provoking paradoxical herniation by lumbar puncture after decompressive craniectomy seems low in the 1st month when the condition that prompted the decompression, such as brain swelling associated with stroke or trauma, will require time to resolve. Although the numbers are small, we found that 1 month after their head trauma, 2 of 10 patients experienced paradoxical herniation provoked by a lumbar puncture. In this setting, lumbar punctures should be avoided if possible, especially if the patient has enlarged ventricles or lacks a bulging skin flap. When the condition that prompted the decompression is addressed with surgery, such as the mass of a brain tumor, the risk of lumbar puncture and lumbar drain provoking paradoxical herniation can be more immediate. Similarly, patients who undergo decompressive craniectomy and develop hydrocephalus are at risk for developing paradoxical herniation at variable times after the placement of a ventriculoperitoneal shunt, depending on the amount of fluid drained and the stage of brain swelling. In our series, 3 of 10 patients so treated deteriorated. Hence, ventriculoperitoneal shunting should be used cautiously in this setting and special attention devoted to clinical response, amount of fluid drained, and appearance of the skin flap. Cranioplasty eliminates the risk of paradoxical herniation and should be considered sooner rather than later in such patients.

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References


Author Contributions

Conception and design: all authors. Acquisition of data: Creutzfeldt, Longstreth. Analysis and interpretation of data: Creutzfeldt, Longstreth. Drafting the article: all authors. Reviewed submitted version of manuscript: all authors. Approved the final version of the manuscript on behalf of all authors: Creutzfeldt. Statistical analysis: Longstreth. Administrative/technical/material support: Longstreth. Study supervision: Vilela, Longstreth.

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