Syndrome of the trephined

- Uniportal Full Endoscopic Posterolateral Transforaminal Lumbar Interbody Fusion
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Grant and Norcross in 1939 first described the syndrome of trephined or paradoxical herniation of brain [1)]

The “Motor Trephine Syndrome (MTS)” also known as the “Sunken brain and Scalp Flap Syndrome” or the “Sinking Skin Flap Syndrome (SSFS)” or the “Syndrome of the trephined” is an unusual syndrome in which neurological deterioration occurs following removal of a large skull bone flap.

The conversion of the cranium from a “closed box” to an “open box” alters the barometric pressure, cerebrospinal fluid (CSF), and cerebral blood flow (CBF) and may or may not lead to syndrome of the trephined [2)].

In 1977 Yamura and Makino coined the term “syndrome of the sunken skin flap” to describe the neurological symptoms due to a craniectomy defect [3)].

Certain patients are particularly susceptible to the presence of a large skull defect. The term “Neurological Susceptibility to a Skull Defect” (NSSD) is therefore suggested as a blanket term to describe any neurological change attributable to the absence of cranial coverage [4)].

The neurological deterioration can be exacerbated or precipitated by CSF diversion procedures like a ventriculoperitoneal shunt.

If one considers Fodstad et al.’s remarks about the effects of atmospheric pressure in patients with skull defects, it is also conceivable that in patients with craniectomies too close to the midline, the sagittal sinus is more prone to collapse by atmospheric pressure. In this situation the normal CSF to sagittal sinus pressure gradient would be lost, leading to poor CSF absorption and thus an increase in extra-axial fluid collections and ventriculomegaly; cranioplasty would reverse this effect [5)].

Epidemiology

Despite the early recognition of neurological symptoms directly linked to craniectomy, the description of this syndrome has often relied on a small series or single clinical case reports. It may be more common than had been previously appreciated [6].

Etiology

The exact cause of the Syndrome of the Trephined is not fully understood, but several potential factors are believed to contribute to its development:

Loss of Skull Integrity: The removal of a portion of the skull disrupts the protective barrier around the brain. This may lead to changes in the pressure dynamics within the cranial cavity. Without the normal skull structure, the brain is less supported, and the absence of the bone may create abnormal fluid or pressure changes that affect brain function.

Increased Intracranial Pressure (ICP) Variability: The craniectomy and the resulting defect in the skull can lead to changes in intracranial pressure regulation. These variations in ICP, particularly after the surgical procedure when the brain is exposed, may contribute to the symptoms associated with the syndrome.

Disruption of Blood Flow: The removal of part of the skull can impact the normal flow of blood within the brain, leading to vascular issues that might result in ischemia or other circulatory disturbances, which can trigger neurological symptoms.

Brain Herniation or Shifting: After craniectomy, the brain may shift or herniate due to the absence of skull support, leading to stretching of the brain tissue or blood vessels, which can cause symptoms such as headaches, changes in consciousness, or neurological deficits.

Altered Cerebrospinal Fluid (CSF) Dynamics: The CSF system plays an important role in maintaining intracranial pressure and cushioning the brain. The craniectomy alters CSF flow, potentially contributing to intracranial hypotension or other disturbances that may present as part of the syndrome.

Psychological and Cognitive Factors: The dramatic change in the head's structure and the neurological symptoms following a craniectomy can result in psychological effects, such as depression or anxiety, which might also contribute to the overall clinical picture of the Syndrome of the Trephined.

Neuroplasticity and Compensatory Mechanisms: After the surgery, the brain might struggle to adapt to the new, altered environment inside the skull. This adjustment period may lead to cognitive and motor dysfunctions, as the brain tries to compensate for the loss of cranial integrity and the changes in its biomechanical environment.

Pathophysiology

Various factors like stretching of the dura and underlying cortex due to the atmospheric pressure,

cicatrical changes occurring between the cortex, dura and the skin exerting pressure on the skull contents, and impairment of the venous return due to the atmospheric pressure acting on the region of skull defect with a resultant increase in the local external pressure have been implicated in the pathophysiology of the “syndrome of the trephined” [7) 8) 9)].

Clinical features

Syndrome of the trephined clinical features

Treatment

see Cranioplasty for Syndrome of the trephined.

Systematic Reviews

Mustroph et al. performed a systematic review of the literature on SoT with a focus on reconstructive implications. Search terms “syndrome of the trephined” and “sunken flap syndrome” were applied to PubMed to identify primary studies through October 2021. The full-text review yielded 11 articles discussing SoT and reconstructive techniques or implications with 56 patients undergoing cranial reconstruction. The average age of the patients was 41.8±9.5 years. Sixty-three percent of the patients were male. The most common indication for craniectomy was traumatic brain injury (43%), followed by tumor resection (23%), intracerebral hemorrhage (11%), and aneurysmal subarachnoid hemorrhage (2%). Patients most commonly suffered from motor deficits (52%), decreased wakefulness (30%), depression or anxiety (21%), speech deficits (16%), headache (16%), and cognitive difficulties (2%). The time until the presentation of symptoms following decompression was 4.4±8.9 months. Patients typically underwent cranioplasty with polyetheretherketone (48%), titanium mesh (21%), split-thickness calvarial bone (16%), full-thickness calvarial bone (14%), or split-thickness rib graft (4%). Eight percent of patients required free tissue transfer for soft tissue coverage. Traumatic Brain Injury (TBI) was a risk factor for the development of SoT when adjusting for age and sex (odds ratio: 8.2, 95% confidence interval: 1.2-8.9). No difference significant difference was observed between length until initial improvement of neurological symptoms following autologous versus allograft reconstruction (P=0.47). SoT can be a neurologically devastating complication of decompressive craniectomy which can resolve following urgent cranioplasty. Familiarity with this syndrome and its reconstructive implications is critical for the plastic surgery provider, who may be called upon to assist with these urgent cases [10)].

2016

Electronic searches of PubMed, MEDLINE, Web of Knowledge, and PsycINFO databases used the key words “syndrome of the trephined” and “sinking skin flap.” Non-English-language and duplicate articles were eliminated. Title and abstract reviews were selected for relevance. Full-text reviews were selected for articles providing individual characteristics of SoT patients.

A review identified that SoT most often occurs in male patients (60%) at 5.1 ± 10.8 months after

craniectomy for neurotrauma (38%). The average reported craniectomy is 88.3 ± 34.4 cm and usually exists with a “sunken skin flap” (93%). Symptoms most commonly include motor, cognitive, and language deficits (57%, 41%, 28%, respectively), with improvement after cranioplasty within 3.8 ± 3.9 days. Functional independence with activities of daily living is achieved by 54.9% of patients after 2.9 ± 3.4 months of rehabilitation. However, evaluation of SoT is inconsistent, with only 53% of reports documenting objective studies.

SoT is a variable phenomenon associated with a prolonged time to cranioplasty. Due to current weaknesses in objectivity, Ashayeri et al., hypothesize that SoT is often underdiagnosed and recommend a multifaceted approach for consistent evaluation.

SoT is a serious complication that lacks exact characterization and deserves future investigation. Improved understanding and recognition have important implications for early intervention and patient outcomes [11)].

2015

Annan et al. selected the references for this review by searching PubMed, focusing on articles published prior to June 2013 and using references from relevant articles.

They used the following search terms: 'trephined syndrome', 'syndrome of the trephined', 'Sinking skin flap', and 'sinking skin flap syndrome'. There were no language restrictions. The final reference list was generated on the basis of its relevance to the topics covered in this review.

Clinicians need to be aware of sinking skin flap syndrome and to look for abnormal neurological developments in patients with craniectomy in order to avoid unnecessary testing and to prevent its occurrence. Accordingly, cranioplasty can be undertaken as soon as necessary [12)].

Case series

Twenty-seven patients (63% with SSS). The most common indication for DC was traumatic brain injury: 48.15%. The p50 diameter of DC was 12.8 cm for patients with SSS and 11.1 cm for patients without (Z score = 0.32). DC area was 81.5 cm2 for patients with SSS and 71.43 cm2 for patients without the syndrome (Z score = 0.61). According to the shape of the craniectomy flap, we classified our patients as: «same level» (51.8%), «sunken» (25.9%), and «extracranial herniation» (14.8%). Two patients (7.4%) had paradoxical herniation. Midline deviation was present in 12 (70.6%) patients with SSS. The 3rd ventricle volume average was 1.2 cc for patients with SSS versus 2.35 cc for patients without (Z score = 0.04). About 94.11% of patients (16 out of 17) clearly improved after the replacement of the cranial defect.

Low 3rd ventricle volumes had a good relation with SSS. The presence of a sunken flap does not guarantee SSS per se and we propose the following radiologic description: A = sunken, B = same level, C = extracranial herniation, and D = paradoxical. Replacement of the skull defect is the main treatment [13)].

1984

Forty patients with cranial bone defects after craniectomy underwent extensive cerebrospinal fluid (CSF) hydrodynamic investigations by means of a CSF infusion test before and after cranioplasty. The results of these investigations were related to the clinical signs of the patients before and after cranioplasty and to the size and location of the skull bone defect. Twenty-two patients were considered to have “the syndrome of the trephined” (ST). The remaining patients were either free of symptoms or had symptoms not related to ST. CSF hydrodynamic variables that were changed before and normalized after cranioplasty include the following: Resting pressure, sagittal sinus pressure, buffer volume, elastance at resting pressure and pulse variations at resting pressure. The changes were statistically significant mainly in ST patients who were also relieved of their symptoms after cranioplasty [14].

Case reports

Syndrome of the trephined case reports.

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