Syndrome of the trephined

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 1).

In 1977 Yamura and Makino coined the term “syndrome of the sunken skin flap” to describe the neurological symptoms due to a craniectomy defect 2).

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 3).

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 4).

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” 5) 6) 7).

Clinical features

Syndrome of the trephined clinical features

2015

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 extraaxial fluid collections and ventriculomegaly; cranioplasty would reverse this effect 8).

A case report of syndrome of the trephined revealed by vertical diplopia 9).
An extreme syndrome of the trephined after decompressive craniectomy is reported by Bijlenga et al. The most extensive clinical syndrome observed was established over 4 weeks and consisted of bradypsychia, dysartria, and limb rigidity with equine varus feet predominating on the right. The syndrome was aggravated when the patient was sitting with the sequential appearance over minutes of a typical parkinsonian levodopa-resistant tremor starting on the right side, extending to all four limbs, followed by diplopia resulting from a left abducens nerve palsy followed by a left-sided mydriasis. All signs recovered within 1-2 h after horizontalisation. It was correlated with an orthostatic progressive sinking of the skin flap, MRI and CT scan mesodiencephalic distortion without evidence of parenchymal lesion. Brain stem auditory evoked potential wave III latency increases were observed on the right side on verticalisation of the patient. EEG exploration excluded any epileptic activity. Symptoms were fully recovered within 2 days after cranioplasty was performed. The cranioplasty had to be removed twice due to infection. Bradypsychia, speech fluency, limb rigidity and tremor reappeared within a week after removal of the prosthesis. While waiting for sterilisation of the operative site, the symptoms were successfully prevented by a custom-made transparent suction-cup helmet before completion of cranioplasty 10).

**Treatment**

see Cranioplasty for Syndrome of the trephined.

**Reviews**

**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 \(^\text{12}\).

**Case series**

**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 \(^\text{13}\).

**Case reports**

**2015**

A 52 year old male suffered severe head injury in a road traffic accident and underwent a craniectomy and contusectomy of the left Fronto-Temporo-Parietal (FTP) region for treatment of Acute Subdural hematoma (SDH) as well as hemorrhagic and non-hemorrhagic contusions of the brain with severe mass effect. On recovery from this acute event he was bed bound, on tracheostomy, his GCS was E4VTrM4 with residual right sided hemiparesis. Three months later, he developed Hydrocephalus for which a Right Ventriculo-Peritoneal (V-P) shunt was performed. Following this procedure, severe depression of the skin/scalp flap occurred and the neurological recovery was not as expected. He was diagnosed as a case of “Syndrome of the trephined”. An immediate Cranioplasty was performed, on the third month following the craniectomy procedure, in an attempt to resolve the rapidly deteriorating neurological status of the patient.

In the case presented, following the early Cranioplasty which was performed within three months of the initial craniectomy, the patient's neurological condition and cognitive functions showed a remarkable, immediate and dramatic improvement. The early Cranioplastic repair led to a remarkable clinical recovery of the patient, with improvement in the cognitive behavior and motor deficit with a rapid reversal of the sensorimotor paresis, reflecting an improvement in brain perfusion \(^\text{14}\).

**2012**

Kwon et al., report a case of a patient with sinking skin flap syndrome who suffered from reperfusion injury after cranioplasty \(^\text{15}\).

**2009**

A 77-year-old male patient with an acute subdural hematoma was treated using a hemicraniectomy and evacuation of the hematoma. On the 9th postoperative day there was deterioration in sensorium associated with a sunken scalp flap and worsening midline shift on CT. A significant improvement in
sensorium and a filling up of the scalp flap occurred after maintaining the patient's head in a dependent position. The patient subsequently made an excellent recovery following replacement of the bone flap.\textsuperscript{16}

2004

A 45-year-old lady underwent right fronto-parietal craniotomy and subtotal excision of a parasagittal meningioma. Bone flap was not replaced as it was infiltrated by the tumor. In the postoperative period she developed episodes of altered sensorium associated with worsening of left hemiparesis and a sunken scalp at the site of bone defect. Computed tomography (CT) of brain showed sunken scalp flap in the right fronto-parietal region with compression of the underlying brain. A diagnosis of syndrome of the trephined was considered and her symptoms improved with cranioplasty.\textsuperscript{17}


