Neurological Impairment due to a Large Skull Defect: Implications for Neurorehabilitation

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Abstract

Given the continued use of decompressive craniectomy in the management of neurological emergencies recognition of complications is important in order for patients to gain maximal benefit during rehabilitation. One complication that has received relatively little attention is the neurological dysfunction that can occur due to distortion of the brain under the scalp as cerebral oedema subsides.

The neurological deterioration that can occur can take many forms and this is probably due to a multifactorial pathophysiology. Recognition of this condition is important if delays in the rehabilitation process are to be avoided. This review discusses the historical background, possible pathophysiological mechanisms, clinical incidence and implications for healthcare workers involved in neurorehabilitation.

Keywords: Cranietomy; Neurological dysfunction; Cerebral oedema; Neurorehabilitation

Introduction

There continues to be a considerable amount of interest in the use of decompressive craniectomy in the management of neurological emergencies [1]. The procedure itself is technically straightforward and involves temporarily removing a large segment of the skull either unilaterally or bifrontally, in order to allow more room into which the injured or ischaemic brain can expand. Its use has been described for a number of pathological conditions, however initially its use was described in the context of either severe traumatic brain injury [2-4] or ischaemic stroke [5,6]. In recent years clinical indications have expanded to include treatment of cerebral swelling in a variety of pathological conditions including subarachnoid haemorrhage [7,8] severe intracranial infection [9,10] dural sinus thrombosis [11,12] and inflammatory conditions [13,14].

Once the cerebral swelling has subsided a cranioplasty procedure is performed in order to replace the bone flap and restore the cranial contour and this usually occurs approximately three to four months after the initial decompressive procedure. Throughout these period patients can face a long and often protracted recovery and are susceptible to a number of complications which can have a significant effect on the rehabilitation process [15]. These include; infection, electrolyte disturbances, seizures and cerebrospinal hydrodynamic disturbances such as hydrocephalus [16].

One complication that has received relatively little attention is the neurological dysfunction that can occur due to the absence of the bone flap and the subsequent distortion of the brain that occurs under the scalp as cerebral swelling subsides. Various terms have been used to describe the wide variety of different neurological manifestations with which this dysfunction can present and until recently these conditions were thought to be relatively uncommon. However it is becoming apparent that a significant number of patients are particularly susceptible to this phenomenon and they may present with subtle functional deficits that may not be appreciated on routine clinical evaluation [17-19]. An appreciation of this phenomenon amongst healthcare workers is important given the pivotal role that rehabilitation plays in the long term outcome of neurotrauma patients and also because there can sometimes be a relative scarcity of rehabilitation resources [20,21]. The aim of this review is to discuss the historical perspectives, proposed pathophysiology and clinical incidence of this phenomenon and thereafter the implications for healthcare workers involved in the neurorehabilitation process.

Neurological Impairment Secondary to a Large Skull Defect – Historical Perspectives

There have been numerous terms applied to describe this condition the first of which was ‘Syndrome of the trephined’ and this was coined by Grant and Norcross in 1939. Their initial description was of subjective complaints from some individuals with a large skull defect and these were documented as; dizziness, undue fatigability, vague discomfort at the site of the defect, a feeling of apprehension and insecurity, mental depression and intolerance to vibration. Although they have been credited with the initial term and description they were by no means the first to notice these clinical symptoms. Indeed they made no such claim and their original article cited 137 articles that dealt with reconstruction of cranial defects many of which described similar clinical changes [22]. Thereafter a number of terms have been suggested that describe what a different manifestation of the same condition is in effect,. Grantham used the term “the post traumatic syndrome” to describe similar subjective symptoms to that of “syndrome of the trephined” [23]. Yamaura and Makino used the term “syndrome of the sinking scalp flap” to describe the objective focal neurological deficits that can occur in patients with a hemicraniectomy defect [24]. “Motor trephined syndrome” is another term used to describe objective motor deficits [25].

In the classical descriptions patients who exhibit this type of signs and symptoms, do so after an initial period of improvement following the decompressive surgery. Thereafter as the scalp flap sinks there is a...
period of clinical deterioration and the diagnosis is confirmed when the symptoms resolve or improve following replacement of the bone flap [26].

Unfortunately, despite the numerous terms available, allotting a patient a specific diagnosis can be problematic because patients can present with a wide range of clinical signs and symptoms. There can be little doubt that some patients do present with the classical features, however some patients have merely been noted to clinically improve following cranioplasty [17,18]. In addition, there is considerable overlap between the diagnostic categories. For example a patient may develop a focal deficit such as a hemiparesis and be deemed to be suffering from ‘syndrome of the sinking scalp flap’ or indeed ‘motor trephined syndrome’. However, on closer questioning they may be found to be having postural headaches and other subjective symptoms which would therefore diagnose them with ‘Syndrome of the trephined’. It may actually be unrealistic to apply a single term that covers all clinical presentations, indeed the differing ways in which patients present may be a reflection of what is in fact a multifactorial pathophysiology.

**Neurological Impairment Secondary to a Large Skull Defect – Pathophysiology**

The underlying pathophysiology responsible for the various neurological manifestations has yet to be established however, a number of theories have been proposed including; direct effects of atmospheric air on the brain, alterations in CSF hydrodynamics and changes in cerebral blood flow.

**Direct Effects of Atmospheric Air on the Brain**

In normal physiological circumstances the brain can float in supportive CSF and fills the confines of the cranial cavity. Once the “closed box” or skull has been opened the principles of the Monroe-Kellie doctrine no longer apply and the brain will be exposed to atmospheric pressure causing distortion not only of the cerebral cortex but also other intracranial structures such as the dura and cranial nerves (Figure 1). This may be the cause of posture related signs and symptoms such as headache, altered sensorium, cranial nerve palsies and mydriasis [27,28].

**Disturbance of CSF Hydrodynamics Following Decompressive Craniectomy**

In the upright position the intracranial pressure will usually be negative however in patients with a large skull defect the ICP will equalize with atmospheric pressure causing distortion not only of the cerebral cortex but also other intracranial structures such as the dura and cranial nerves (Figure 1). This may be the cause of posture related signs and symptoms such as headache, altered sensorium, cranial nerve palsies and mydriasis [27,28].

**Disturbance in Cerebral Blood Flow and Metabolism**

A number of studies have demonstrated the alterations in cerebral blood flow that can occur under a large skull defect and the subsequent improvement in blood flow that can occur following cranioplasty [29,30].

The pathophysiology underlying this vascular response is unknown but may be due in part to the transmission of atmospheric pressure on to the cerebral vasculature combined with normalisation of CSF compliance and cerebrovascular auto-regulatory function [31].

Overall it would seem most likely that a large skull defect can have numerous effects on the cerebrovascular physiology and CSF hydrodynamics and there will be no single pathophysiological mechanism to account for the wide variety of clinical manifestations reported.

**Neurological Impairment Secondary to a Large Skull Defect – Clinical Incidence**

The true incidence of this clinical condition remains unknown. Earlier reports regarding neurological dysfunction due to a large skull defect have described these manifestations as either rare or uncommon however most publications were either case reports or small retrospective cohort studies. Whilst some of them did describe impressive neurological recoveries there was often no baseline denominator recording the number of patients for whom the cranioplasty had no clinical impact.

More recently a prospective cohort study found an objective improvement in neurological function in four (16%) out of twenty five patients who were assessed a few days before and after cranioplasty however more work on larger case series will be required to determine not only the true incidence but also what factors predispose patients to this condition [1]. These issues may be important when considering the impact that this can have on rehabilitation and also the timing of the cranioplasty procedure.

**Neurological Impairment Secondary to a Large Skull Defect – Implications for Rehabilitation**

The most important implications for rehabilitation is perhaps not only recognising that this condition exists but also appreciating that the degree to which it affects individual patients and the clinical manifestation thereafter can vary considerably. This in itself can present considerable diagnostic difficulty especially when the physical signs and symptoms are subtle. In the first instance any patient who either fails to steadily improve during the rehabilitation process or indeed who deteriorates while awaiting a cranioplasty procedure must be fully investigates. This will allow common problems such as haematological or biochemical anomalies and sepsis to be addressed. Thereafter consideration may be given to early consultation with the neurosurgical team with a view to expediting the cranial reconstruction.

Currently, optimal timing of cranioplasty has not been clearly established. For many years it was suggested that the procedure should be delayed in order to reduce the risk of infection, however recent studies have suggested that early cranioplasty can be safely performed [15]. In view of these findings it would seem logical to replace the bone flap...
flap as soon as clinically possible given that rehabilitation facilities are often a scarce and valuable resource. Every effort must be made to ensure that these resources are deployed appropriately in order to provide as many patients with maximal benefit. Indeed, given the impact that a large skull defect can have on neurological recovery, it has been suggested that intensive neurorehabilitation should not be undertaken until a cranioplasty has been performed [18]. Whilst this may seem a reasonable position to adopt it would mean that those patients not affected by the skull defect would miss out on the potential benefit of early rehabilitation. A more realistic approach would be to highlight the need to recognize the condition in susceptible individuals and it in this regard that all healthcare workers involved in the neurorehabilitation process can contribute.

Conclusions

Patients who have had a decompressive craniectomy face a particularly challenging recovery and all efforts should be made to maximize the potential for neurological recovery. It is becoming increasingly apparent that certain individuals are particularly susceptible to having a large skull defect and it is important that this is recognised so that consideration may be given for early cranioplasty in order to minimise any disruption to the rehabilitation process.

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