A Systematic Review and Illustrative Case of Post-Decompressive Craniectomy Syndrome Following Traumatic Brain Injury

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ABSTRACT

Post-decompressive craniectomy syndrome (PDCS) is a complication following decompressive craniectomy (DC). PDCS or also known as trephine or sunken skin flap syndrome has an indirect relationship with traumatic brain injury (TBI). The mechanism of PDCS is not yet fully understood and the clinical manifestations are diverse, causing PDCS to often be underdiagnosed. In this study, the authors aim to create a systematic review of PDCS following TBI including a discussion of incidence, clinical and radiological manifestations, management and outcome. This systematic review is conducted based on the Preferred Reporting Items for Systematic Review and Meta-Analysis (PRISMA) guideline. The literature search included electronic databases PubMed, Cochrane, JNS and EMBASE. All studies included were available in English and full-text format. In this research, 42 case reports were obtained. The population was predominantly male (72%) with a mean population age of 44.7 ± 17.3 years. The mean interval for onset and cranioplasty procedure was 80.17 ± 77.34 days and 92.05 ± 77.06, respectively. The most common clinical manifestations were sunken skin flap in the defect area (74%) and decreased consciousness (64%). Paradoxical herniation (74%) was the most common radiological manifestation. There was no connection between the occurrence of PDCS and the size of the defect. Cranioplasty remains the mainstay of management with clinical improvement in 96% of cases. PDCS should be suspected in every patient with symptoms of new neurological deficits after DC. Early management must be carried out to prevent further deterioration.

Keywords: Post-decompressive craniectomy syndrome, complication, decompressive craniectomy, cranioplasty
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INTRODUCTION

Post-decompressive craniectomy syndrome (PDCS) or trephine or sunken skin flap syndrome is a complication of decompressive craniectomy (DC) which describes new neurological deficits due to cortical dysfunction caused by brain compression in the defect area.[1–3] The incidence of this syndrome is estimated at 10% to 20% in patients after DC procedures.[4,5] Abnormalities in normal anatomy and physiology may cause a range of symptoms, including mental status abnormalities, hemodynamic disturbances, and neurological impairments.[6,7] The clinical and radiological characteristics presenting with PDCS are often atypical, making it important to consider these complications in each post-DC patient.[8,9]

One of the most common causes of increased intracranial pressure is traumatic brain injury (TBI).[10,11] Brain edema

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due to TBI causes intracranial hypertension.[12,13] DC is a procedure in the field of neurosurgery where skull bones are removed with the aim of reducing intracranial pressure.[14] The rationale for carrying out DC is the closed box concept of the intracranial cavity based on the Monro-Kellie doctrine.[15] DC provides the potential space of the cranial cavity and allows the expansion of the brain.[16]

DC has the effect of causing the cortex to become vulnerable to external pressure due to a skull defect.[17] Cranioplasty is a procedure to close cranial defects and is still considered the primary management to this day. Cranioplasty is performed to prevent or eliminate collapse of the brain parenchyma and the brain remains mechanically protected. Closure of bone defects is necessary to prevent differences in atmospheric pressure pressing on the cortex in the defect area.[18] Several concepts have been put up to attempt to explain the physiology of this disease, but the explanation is still elusive.[19,20]

The diverse manifestation of PDCS and its unclear mechanisms may contribute to its underdiagnosis and under-representation in the neurosurgical literature, resulting in a lack of awareness of this relatively common disorder within the neurosurgical community. Thus, we conducted a systematic review to discuss PDCS following TBI, which currently does not have much literature discussing this matter.

MATERIALS AND METHODS

Study design and literature search strategy

This systematic review aimed to review the incidence and complications of PDCS. The search strategy for journals was carried out referring to the Preferred Reporting Items for Systematic Review and Meta-Analysis Protocol (PRISMA) guideline (Fig 1). Journal data was collected through several databases such as PubMed, Cochrane, JNS, EMBASE. The articles searched had no limitations on the year published and used the keywords "Trephine Syndrome" OR "Sunken skin flap syndrome" AND "Traumatic brain injury" AND "Craniectomy" AND "Cranioplasty". Authors reviewed the reference lists of all relevant studies including published studies. We excluded all unpublished studies or articles-in-press.

The definition of the syndrome was (1) a neurological deficit that usually appears after DC, (2) new complications that appear not as a sequel to the initial lesion, and (3) clinical improvement after cranioplasty. Subjects were patients who underwent a traumatic DC procedure with age >16 years. Exclusion criteria were subjects with age < 16 years and the cause of craniectomy other than trauma.

Data collection and synthesis process

Articles were accepted for this review if they were written in English or Indonesian and had no publication year restrictions. The data studied included sex, age, timing of first symptom and cranioplasty, sign and manifestation, defect area, outcome and bias. Statistical averages and percentages for all populations and characteristics were examined and described descriptively.

Illustrative case

A 35-year-old male patient presented with severe headache, came to our emergency department at Dr. Soetomo General Academic Hospital, Surabaya, Indonesia. He had a history of decompressive craniectomy procedure 3 months prior due to traumatic intracerebral haemorrhage and skull depressed fracture. His symptoms worsened 1 month before admission. Physical examination showed a visible sunken skin flap in the defect area. Brain computed tomography (CT) scan revealed paradoxical herniation in the defect area (Fig 2). The patient underwent cranioplasty. The patient had an uneventful postoperative course and was discharged with no headaches, emesis, or new neurological deficits.

RESULTS

The literature search yielded a total of 42 studies. The research flow diagram can be seen in Fig 1. The databases searched included PubMed, Cochrane, EMBASE, and JNS. The search was conducted in January 2024. After removing duplicates, we removed 489 articles in the abstract review and a further 288 articles in the full-text review. Of the remaining studies, we continued with a systematic review for 42 eligible studies with a total of 47 patients. All studies founded during the strategy process were either a case report or a case series. A case report or case series is a study that describes the course of a patient's condition descriptively. This is differentiated from a quantitative study in which analysis and comparisons are carried out between outcomes or treatments. Authors found no other quantitative studies such as cohort, case control or cross sectional studies.

Sex and age

The sex distribution is dominated by male (72%) compared to female (28%). The population's average age was 44.7 ± 17.3 years (Table 1).

Timing of onset and cranioplasty

The mean interval for symptoms to appear after the decompressive craniectomy procedure was 80.17 ± 77.34 days. The mean interval value for the cranioplasty

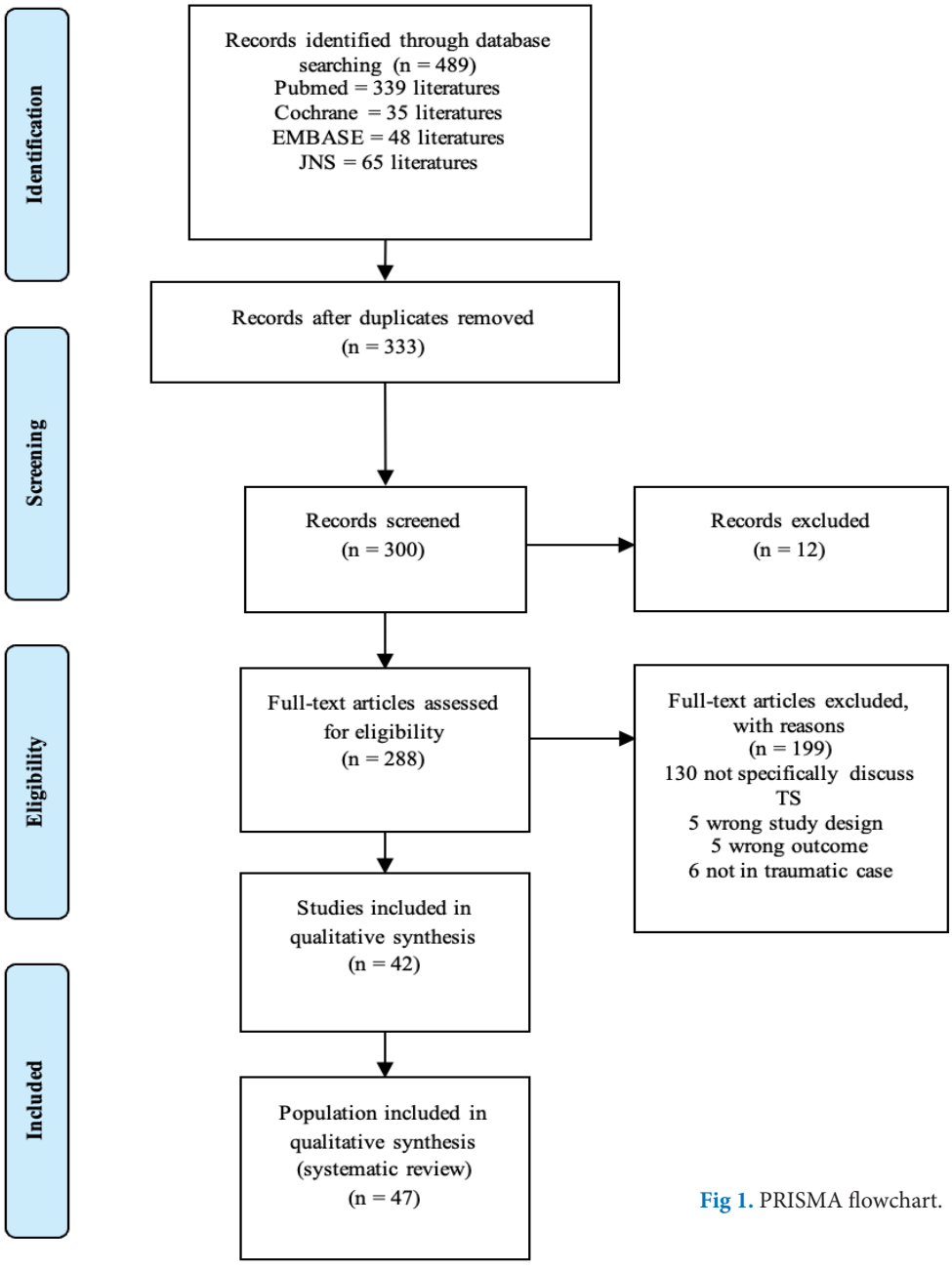

Fig 1. PRISMA flowchart.

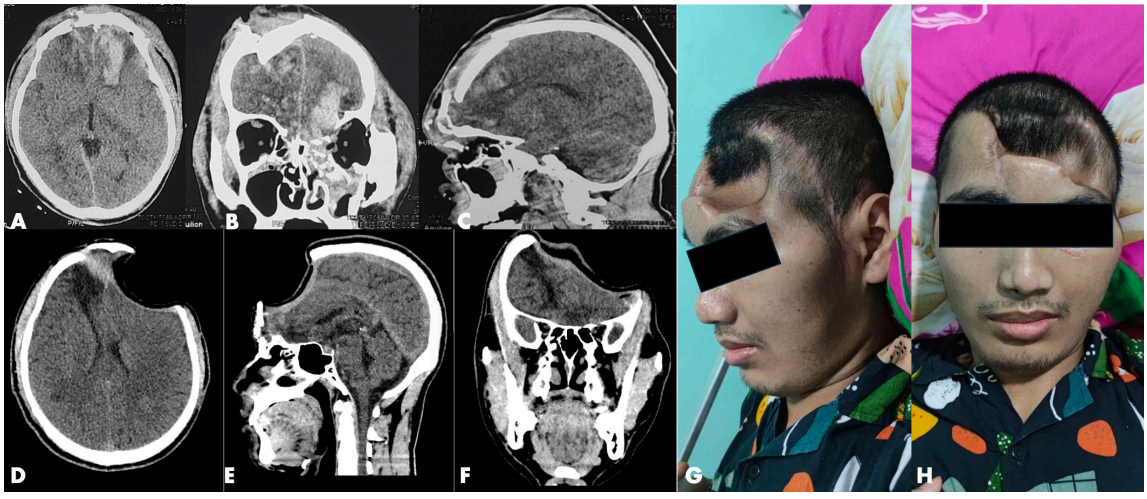

Fig 2. Preoperative CT revealed intracerebral haemorrhage and depressed fracture (A, B, and C). Radiological (D, E, and F) and clinical (G and H) images show features of the syndrome in the defect area after 3 months post-DC procedure.

TABLE 1. Demographic data.

Characteristic		No. (%) of Cases
Sex (n)	Male (%)	34 (72)
	Female (%)	13 (28)
Age (year)		44.7 ± 17.3
Timing from DC to onset (days)		80.17 ± 77.34
Timing from DC to cranioplasty (days)		92.05 ± 77.06

procedure was 92.05 ± 77.06 days after the decompressive craniectomy procedure (Table 1).

Clinical manifestation

The results of clinical manifestations in cases of TS after traumatic decompressive craniectomy are shown in Table 2. The most common clinical manifestations are sunken skin flap in the defect area (74%), and decreased consciousness (64%). Other symptoms include motor weakness (43%), headache (30%), language deficits (23%), worsening of positional symptoms (19%), cognitive deficits (13%), cranial nerve deficits (13%), seizures (10%), psychosomatic (4%), and sensory deficits (2%). Literature analysis reported no cases of mortality.

Radiographic image

The radiological features of the supporting examinations carried out are as shown in Table 3. Paradoxical herniation

TABLE 2. Clinical manifestation.

Clinical feature	No. of Cases	Percentage (%)
Sunken skin flap	35	74
Decreased consciousness	30	64
Motor weakness	20	43
Headache	14	30
Language deficits	11	23
Positional symptoms	9	19
Cognitive deficits	6	13
Cranial nerve deficits	6	13
Seizure	5	10
Psychosomatics	2	4
Sensory deficits	1	2
Mortality	0	0

TABLE 3. Radiographic image.

Radiological feature	No. of Cases	Percentage (%)
Paradoxical herniation	35	74
Hydrocephalus	11	23
Infarction	0	0

was the most common radiological presentation (74%), followed by hydrocephalus (23%). Literature analysis reported no features of infarction.

Defect area

Only 2 studies reported the size of the defect in a population of PDCS cases after traumatic craniectomy decompression, namely 43.6 cm^2 and 110 cm^2.

Outcome

In the majority of cases (n=45) there was clinical improvement after the cranioplasty procedure (96%).

Bias

An analysis was carried out to assess the risk of bias: confounding, selection, information, and reporting bias on 42 pieces of literature that underwent a systematic review. Details of the results of the risk of bias analysis can be seen in Fig 3. Authors assessed the bias based on the modified Cochrane Collaboration tool : Risk of Bias in Non-randomised Studies–of Interventions' (ROBINS-I).[20]

DISCUSSION

Epidemiology

Demographic factors such as age and gender may be risk factors that influence outcomes after craniectomy and cranioplasty. A study conducted by Santander et al., reported that in terms of age and gender, there was no difference between patients with or without PDCS.[21] The majority of the PDCS case population was male, which was also related to the fact that the predominance of the patient population undergoing DC following TBI occurred mostly in male patients.[22] Men were also predominant in this study. Demographic factors such as age and gender may be risk factors that influence outcomes after decompression and cranioplasty. In a cohort study by Sveikata et al in 2021, of the total of 40 patients studied, age and gender had a low p value on the risk of TS after decompression. These findings led to the conclusion that the incidence of PDCS following TBI was not significantly influenced by age or gender.[23]

Onset and clinical manifestation

The lack of established diagnostic criteria makes it difficult to diagnose PDCS patients early. Recent studies show that radiologically, 81% of patients with PDCS have sunken defect area, hydrocephalus, obliteration of ventricle, but the individual diagnostic yield is still low.[23,24] Approximately 50% of PDCS manifest without classic radiological signs such as sunken skin flaps, and more than 80% without paradoxical brain herniation. This study also showed that the majority of clinical manifestations included improvement after the cranioplasty procedure (96%), visible sunken skin flap (74%), and decreased consciousness (64%). Detailed physical and radiological examinations must be carried out to establish the diagnosis with such diverse manifestations.[23,25,26]

A study by Santander, reported that the majority of presentations were motor impairment (82%).[21] Therefore, some researchers recommend screening for cognitive deficits. Previous studies showed that 47% of patients with PDCS had cognitive impairment.[21,27] As the name of this syndrome suggests, sunken defect area was associated with PDCS in 57.14% cases.[30-31] Several studies also confirmed this, but it must also be taken into

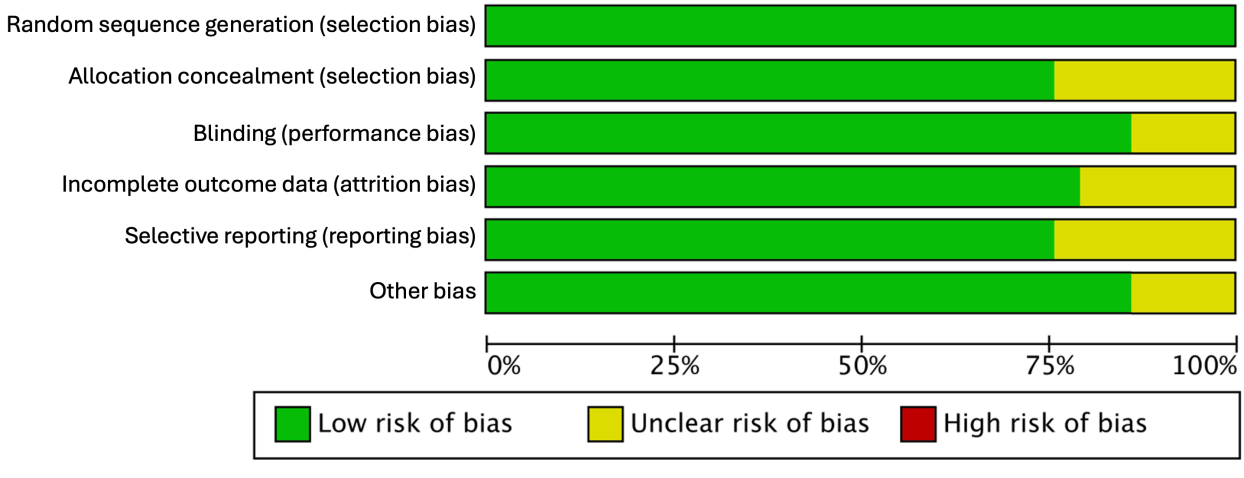

Fig 3. Bias analysis summary. The bias analysis was conducted based on the modified Cochrane Collaboration tool : Risk of Bias in Non-randomised Studies–of Interventions' (ROBINS-I).

account that some patients with sunken defect area had no neurological deficits.[28–30] Therefore, sunken defect areas are not cardinal symptoms of PDCS. This finding also corresponds to the definition used by several other studies, where true PDCS should be suspected when clinical presentation improve after cranioplasty.[31]

Radiological examination

Cerebrospinal fluid, atmospheric pressure, and cerebral blood flow are some of the variables that affect the PDCS occurrence. It is consistent that paradoxical herniation and hydrocephalus were the most common radiological signs in this study. Of these factors, atmospheric pressure is reported to be the main factor that causes compression and damage to cortical tissue in unprotected brain tissue in bone defects, which ultimately causes neurological deficits and then repair after cranioplasty.[32] Imaging with cranial F18-fluorodeoxyglucose positron emission tomography CT (F-18 FDG-PET/CT) is helpful for assessing brain metabolism. After TBI, molecular shifts and inflammation cause disruptions in glucose metabolism.[33]

Defect area

The size of the craniectomy has long been considered a factor in this syndrome. There is no correlation published in the literature, but craniectomies with an area >100 cm^2 may be associated with the incidence of PDCS.[22]

Tarr et al. showed that occurrence of PDCS increased with a craniectomy area of 50 cm^2 or more.[34] Although Sveikata et al showed that there no correlation between the extent of craniectomy in the PDCS and non- PDCS groups, wide craniectomy as an inclusion criterion in the study (mean area 112.8 ± 35.4 cm^2).[23] Our analysis is that PDCS will have a tendency to appear if it exceeds a certain area. So in patients who have extensive DC such as hemicraniectomy, the incidence of PDCS must be watched out for.[34]

Management and outcome

The initial management of PDCS that can be done is positioning from supination to a sitting position in the hope that intracranial pressure will decrease. Cranioplasty should be performed immediately when PDCS is suspected to prevent irreversible recovery of the functional outcome.[35] The incidence of cranioplasty-related complications (such as infection) or the inability to decompress it thoroughly should be monitored.[36]

Cranioplasty remains the primary management of PDCS.[36,37] Previous study reported that the earliest cranioplasty in patients with PDCS can be performed within 18 months after DC. The defect location factor (left, right, front, posterior or bilateral) did not have a significant correlation with the occurrence of PDCS.[29] Cranioplasty management in younger patients shows better postoperative outcomes.[38,39]

In another systematic review study, it was concluded that early cranioplasty (<3 months) does not have significant advantages when compared to late cranioplasty (>6 months). Early cranioplasty could reduce length of stay at hospital, but did not reduce the risk of complications.[40]

Research on cranioplasty after decompressive craniectomy by Safi et al in 2022, which used a sample of 132 patients, stated that the patient's initial level of awareness had a significant influence on post-cranioplasty outcomes.[41] This study analysed multiple logistic regression statistics to identify significant factors related to pre-cranioplasty Glasgow Outcome Scale (GOS), decompression indication, and cranioplasty waiting time. The study concluded that the best Glasgow Coma Scale (GCS) the patient had before the cranioplasty procedure had a significant influence on the outcome (p value 0.001). In 55 patients out of 132 samples who underwent cranioplasty after the decompression procedure had good outcomes with a pre-cranioplasty GOS score of 4-5 (good grade) and poor outcomes in patients with a pre-cranioplasty GOS score of 2-3 (poor grade).[29,34,41]

Limitation of the study

Authors didn't review ongoing trials because there were currently no ongoing trials with this study topic until this study conducted. The author didn't perform meta-analysis study due to two main reasons. First, there was no literature or research reports that randomly compare the effectiveness, outcomes and side events of PDCS. Second, the limited number of samples is due to the fact that all studies are either case reports or case series. The very small number of quantitative studies means that meta-analysis cannot be carried out.

CONCLUSION

The pathomechanism of PDCS is still unclear to date. The atypical clinical manifestations and delayed onset make PDCS difficult to recognize early. Clinicians should be aware of the possibility of PDCS in patients with new post-DC neurologic deficits. Cranioplasty should be planned as soon as the patient meets the criteria for defect closure to prevent further deterioration. Authors hope there will be more quantitative studies in the future so that meta-analysis and research related to PDCS can be carried out, so they will have higher level of evidence and more representative result.

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Conflict of Interest

None

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