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The diagnostic credibility of second impact syndrome: A systematic literature review

Olivia Hebert^a, Kaitlyn Schlueter^a, Meg Hornsby^a, Samantha Van Gorder^a, Suzanne Snodgrass^b, Chad Cook^{c,*}

^a Doctor of Physical Therapy Division, Duke University, United States

^b School of Health Sciences, The University of Newcastle, Australia

^c Doctor of Physical Therapy Division, Department of Orthopaedics, Duke University, United States

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ABSTRACT

Objectives: The purpose of this review was to examine current literature to determine whether or not enough evidence exists to support a World Health Organization (WHO) recognized ICD-10 case definition for Second Impact Syndrome (SIS).

Design: Systematic literature review.

Methods: A sensitive search strategy was developed in order to include all relevant literature. Data were extracted and placed into a diagnostic framework constructed based upon previously accepted ICD-10 diagnoses. The quality of included studies was assessed using a checklist developed specifically for case reports.

Results: The literature search yielded 338 articles. After duplicates were removed, the remaining 222 articles were screened. Seventy-five articles were assessed for full-text eligibility, which resulted in eight case studies appropriate for this review. Significant information regarding imaging and confirmed signs and symptoms is mixed or absent. Information exists to support possible at-risk populations, signs and symptoms.

Conclusions: At present, there lacks a unique presentation scheme of SIS to support a standardized WHO case definition. Furthermore, future studies are needed to better understand and define at risk populations, diagnostic signs and symptoms, and the multisystem consequences of Second Impact Syndrome. © 2016 Sports Medicine Australia. Published by Elsevier Ltd. All rights reserved.

1. Introduction

Concussion is described as a physiological process seen acutely following a mild traumatic brain injury that results in transient functional impairment.¹ The clinical presentation, prognosis, and complication rate varies markedly among individuals suffering concussion. The majority (80–90%) of concussions resolve in a short, 7–10 day period, although the recovery time frame may be longer in children and adolescents.² In some cases, concussion symptoms are prolonged and can lead to a condition known as post-concussion syndrome. Post-concussion syndrome is a complex disorder in which various symptoms such as headache, fatigue, dizziness, and concentration problems associated with the original diagnosis of concussion, are prolonged beyond the point of expected recovery.^{3,4}

* Corresponding author. E-mail address: chad.cook@duke.edu (C. Cook).

Second Impact Syndrome (SIS) is a hypothetical diagnosis which is said to occur when an individual suffers a second head injury before symptoms from a first head injury have resolved.^{5,6} Clinically, SIS has been described uniquely by a second head impact and subsequent emergent signs/symptoms. It is stated that the condition is potentially critical, with a high reported incidence of death and disability.^{5,7} Initially, the individual/athlete who suffers a second impact may appear stunned but have no loss of consciousness, often demonstrating the ability to walk off the field independently before rapidly deteriorating. As time progresses, individuals suffering from SIS may appear semi-comatose with rapidly dilating pupils, suffer loss of consciousness, or present with respiratory failure. During emergent triage, diagnostic imaging typically identifies massive cerebral swelling. The swelling that occurs after a second impact, during a period of heightened brain vulnerability, is thought to be the result of disordered autoregulation of the blood supply to the brain resulting in increased intracranial pressures.^{8,9} It has been suggested that outcomes after a second head injury are poor, with a mortality rate of 50% and a morbidity of 100%.¹⁰

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At present, there remains controversy surrounding the parameters of diagnosis of SIS and mere existence as a condition. SIS is not mentioned in the 2013 Consensus statement on Concussion in Sport (Zurich panel).² Further, concussion (S06.0) and post-concussion syndrome (F07.81) are recognized diagnoses, each having its own World Health Organization (WHO) derived ICD-10 code. SIS lacks a unique ICD-10 code (a code does exist for subsequent concussion) and lacks a definitive clinical case definition within WHO. All ICD-10 codes are copyrighted and published by the WHO, and are used for classification and collection of morbidity and mortality data. ICD-10 codes function as a common diagnostic language worldwide to facilitate effective communication amongst healthcare providers.¹¹ Periodic updates regarding the establishment of new diagnostic codes are the responsibility of the Mortality Reference Group (MRG) and the Updating and Revision Committee (URC) of WHO.¹² To create a case definition for a disease, WHO requires significant information on signs, symptoms, diagnostic guidelines, and at risk populations to create a criteria for assessing a diagnosis.

Most resources that have discussed SIS are secondary sources or position papers, and to our knowledge, there are no contemporary systematic reviews of the literature that examine the evidence to support or refute the signs, symptoms, diagnostic guidelines and at-risk populations for this purported diagnosis. A 1998 review by McCrory and Berkovic¹³ reviewed proposed cases of SIS and compared them to a four part criteria to determine if any of the cases could be uniquely defined as SIS. Of the seventeen cases that were examined only 5/17 were considered to be *probable* cases of diagnosed SIS, and 0/17 were found to be *definitive* cases of SIS. Of these 17 cases, 13 reported sport-related catastrophic brain injury associated with unexplained cerebral swelling, yet this adverse event was typically *not* associated with a second impact.¹⁴

The purpose of this review was to determine whether or not evidence in the current literature supports a unique case definition for SIS, based on a format consistent with WHO, ICD-10 clinical case definitions. This systematic review will investigate the published evidence for SIS to determine support for this diagnosis. With criteria for diagnosing and identifying at risk populations, clinicians will better be able to protect individuals in these populations who have sustained a concussion, from further injury or mortality.

2. Methods

This systematic review used the Preferred Reporting Items for Systematic Reviews and Meta-Analyses (PRISMA) guidelines during the search and reporting phase of the research process. The PRISMA statement includes a 27-item checklist designed to improve reporting of systematic reviews and meta-analyses.¹⁵

The following were the inclusion criteria for the study: (1) English language, (2) publications of any date; (3) primary sourced articles (defined as an original material that has not been filtered through interpretation or evaluation by a second party); (4) casebased reports or case series, specifically implying a condition of SIS; (5) article that lends support toward a pathoanatomical process related to SIS; and (6) accepted outcome metrics required for the composition of a WHO recognized case definition, including, (a) human studies only, (b) physiological signs/symptoms that describe the pathophysiology of SIS (autoregulation, neurological and biological markers, post-death imaging), (c) time-related phenomena, and (d) risk components (including at risk populations). Papers were excluded if they failed to mention second impact syndrome (SIS) directly, a subsequent head injury, or a repetitive or secondary head injury. Four databases (PubMed, CINAHL, SPORTDiscus, and EMBASE) were searched systematically from the earliest available date through January 2015. The search strategy was created with assistance from a biomedical librarian. A sensitive search strategy was used in order to capture any literature that would be relevant in the creation of a WHO characterized ICD-10 case definition. A keyword search was conducted using "second impact syndrome" OR "second-impact syndrome" OR "second impact". The search yield was imported into Zotero for OS X. Zotero is a web-based organization tool allowing for consolidation of literature searches from multiple databases, elimination of duplicate articles, and precise tracking for construction of a PRISMA flow sheet.

Titles and abstracts were first screened independently by two reviewers using previously mentioned inclusion and exclusion criteria. The full texts of the remaining articles were then obtained and screened independently by two reviewers. Differences in opinion between reviewers were discussed until a consensus was reached. Citation tracking using Zotero for OS X and reference checking of the included articles was also performed independently by two reviewers.

World Health Organization Case-Definition Structure: A diagnostic framework addressing the WHO's required criteria for a recognized diagnosis was constructed using the ICD-10 code for post-concussion syndrome as a template. Four broad categories are required to satisfy ICD-10 diagnostic criteria: (1) signs and symptoms, (2) diagnostic guidelines, (3) differential diagnoses, and (4) populations at risk/confirmed cases.

Signs and symptoms were divided into those associated with a suspected case, a probable case and a confirmed case. Diagnostic guidelines included objective information such as blood work and imaging, that are necessary to triage and confirm a state of heightened brain vulnerability in "at risk" populations. A list of possible differential diagnoses was constructed, and included concussion, post-concussion syndrome, epidural hematoma and malignant brain edema. All differential diagnoses resulted from head trauma and/or cerebral swelling.

Data were extracted from result-based components of included case studies and tabulated into a PEOT – Population, Exposure, Outcome, and Type of Study (Table 1). Result-based components of a case study included the examination and evaluation sections of the study. Information was extracted based on support for pathoanatomical signs and symptoms, neuroimaging findings and populations at risk for SIS (Table 2).

One author performed data extraction from included articles. Articles were re-read and cross-examined with the created criteria for a WHO recognized ICD-10 clinical case definition. Appropriate data were dropped into corresponding categories of the WHO case framework, and included a citation of the original source for reference. Conflicting data, defined as different results from different studies, was included in the WHO framework but marked with an asterisk (*) to denote controversy. A second team member reviewed the WHO framework in reference to the included articles to ensure no relevant data were excluded.

Quality assessment was performed using a critical appraisal tool developed specifically for case reports, which is endorsed by the Oxford Centre for Evidence Medicine, checklists of the Dutch Cochrane Centre, BMJ editor's checklists and the checklists of the Evidence for Policy and Practice Information and Coordinating Centre.¹⁶ The tool¹⁶ was originally adapted from Combie's Pocket Guide to Critical Appraisal, and includes 10 items scored as 'yes', 'cannot tell', or 'no'. Items involve scoring of clarity, appropriate methodology, representation, reliability, credibility, appropriateness of conclusions, and transferability. One author reviewed and scored all papers using this tool.

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