Management and Mid-Term Outcome After “Real SCIWORA” in Children and Adolescents

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Abstract

Study Design: Retrospective analysis.
Objective: The SCIWORA Syndrome (Spinal Cord Injury Without Radiographic Abnormalities) is a rare but potentially severe injury with a peak in childhood and adolescence. With a better understanding of injury patterns and advances in MRI, there is ongoing discussion regarding the “Real SCIWORA” syndrome, a clinical picture of neurologic deficits on clinical examination but absence of radiographic pathologies even on MRI. The purpose of this study was to evaluate mid-term clinical outcome and the psychological impact of the “Real SCIWORA.”

Methods: In this retrospective analysis, we evaluated 32 patients treated for “Real SCIWORA” between 2007-2019. Inclusion criteria were: neurologic deficit after trauma, no other cerebral or skeletal injury and a lack of pathological findings in spinal MRI. All patients were followed until complete recovery from initial symptoms. 25/32 patients were re-evaluated after 6.9 years (1-14 years) using the Oswestry Disability Index, the Frankel Score, the EQ-5D score, and the Breslau Short Screening Scale for PTSD.

Results: Initial neurologic presentation ranged from Frankel Grade A-D. All patients recovered neurologically during 1-13 days to a Frankel Grade E. The analysis of HR-QoL revealed no difference between the cohort of SCIWORA patients and the German population norm, Oswestry Disability Index showed only minimal disabilities. 4/25 patients showed signs of PTSD.

Conclusions: The “Real SCIWORA” syndrome is a diagnosis per exclusionem requiring a full spinal MRI to ensure exclusion of structural and potentially serious reasons of the neurologic impairment. Further clinical re-evaluation, psychological support seems to be essential.

Level of Evidence: IV—retrospective study.

Keywords

pediatric and adolescent trauma, management of severe trauma, SCIWORA, patient-reported outcome

In 1982 Pang and Wilberger defined Spinal cord injury without radiographic abnormality or “SCIWORA” as “children with objective signs of myelopathy as a result of trauma, whose plain films of the spine, tomography, and occasionally myelography carried out at the time of admission showed no evidence of skeletal injury or subluxation.” They recommended the use of every radiographic diagnostic known at that time including tomography and flexion-extension radiographs to detect instability. They reported that patients with severe neurology

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do not recover.\textsuperscript{1} But with advances in radiology this definition seems dated. During an era when radiological examination of the spinal cord was limited, it seems likely that the patients treated for SCIWORA included patients with severely injured spinal cords. Our understanding of SCIWORA has evolved as the Magnetic Resonance Imaging (MRI) has improved and become widely available.\textsuperscript{2,3} MRIs enable clinicians to evaluate the intramedullary state of the spinal cord in detail and helps to identify diseases that may require surgical intervention. Also it can distinguish between spinal cord lesions, disc herniations, edema, hematoma, contusions, ischemic injuries and the “Real SCIWORA”—the absence of any spinal abnormality.\textsuperscript{4}

Nonetheless, there are few studies that utilize MRI to rule out more specific diagnoses and use the SCIWORA syndrome as an umbrella term for unspecific injuries. The “Real SCIWORA” syndrome without any radiographic effect on MRI differs from the previously published imprecise definition of SCIWORA syndrome.

The purpose of this study was to evaluate children and adolescent patients who were treated for neurological deficit after trauma, without any pathological finding in spinal MRI to show the mid-term outcome regarding neurological disabilities as well as psychological impact of neurologic impairment.

**Methods**

The study was approved by the Ethics Committee of the University of Regensburg (Institutional Review Board Number 20-1928-101). Informed consent was obtained by all participants. We conducted a retrospective study on “Real SCIWORA” patients between 2007 and 2019 who were admitted to our level I trauma center for traumatic spinal injury.

Inclusion criteria were:

- Post-traumatic sensory-motoric deficit of the upper or lower extremity
- Absence of any sign of post-traumatic effect on a high-resolution MRI scan of the complete spine

Exclusion criteria were:

- Any post-traumatic alteration of the signal within the MRI scan
- Any concomitant injury that might potentially cause a neurologic symptomatic (e.g. traumatic brain injury, fracture or laceration that might go along with a peripheral nerve injury)

The evaluation included all available X-rays, Computed tomography (CT) scans, and a full spinal MRI-scan of every patient. A radiologist and a senior trauma surgeon carried out evaluation. The diagnosis of spinal cord injury was established by a specialist (neurosurgeon or neurologist). All patients were followed until complete recovery of neurologic symptoms. After 6.9 years (1-14 years) we re-evaluated all available patients for an assessment of clinical, functional and psychological mid-term outcome.

The Oswestry Disability Index was applied to quantify disease-specific functional outcome due to back pain. This questionnaire contains: intensity of pain, lifting, ability to care for oneself, ability to walk, ability to sit, sexual function, ability to stand, social life, sleep quality, and ability to travel.\textsuperscript{5}

The Frankel Score was used to assess spinal cord function. It is a 5 grade scale to assess the extent of the neurological/functional deficit into 5 grades: The functional deficit is classified as complete (grade A), complete motor loss (grade B), incomplete motor loss without practical use (grade C), incomplete motor loss, able to ambulate with or without walking aids (grade D), or no neurological deficit/complete recovery (grade E).\textsuperscript{6}

For evaluation of global health-related quality of life (HR-QoL), we used the EQ-5D-3L score. The EQ-5D is a global quality of life questionnaire consisting of a 5-item assessment of the health status regarding mobility, self-care, usual activities, pain/discomfort, and anxiety/depression. The second part of the EQ 5-D consists of a 100 mm global health visual analogue scale (EQ-VAS) ranging from 0-100 points concerning the patient’s assessment of their current global health status. The EQ-5D has been adapted to German and has been validated for a number of health compromising conditions.\textsuperscript{7,8}

For evaluation of possible posttraumatic stress disorder (PTSD), the Breslau Short Screening Scale (BSSS) for PTSD was applied. It contains a seven-symptom screening scale and assesses if the patient: avoids places and people; lost interest in activities; felt isolated; found it hard to have affection for others; had a sense of foreshortened future; had sleep difficulties; or became easily startled. It is well proven to be an efficient way to screen for PTSD.\textsuperscript{9,10}

**Statistics**

Statistical analysis was performed using the software package SPSS (Version 24, SPSS Inc, Chicago, Illinois). Level of significance was defined at \(p < 0.05\) for all tests. Descriptive data are given as frequencies (n) and percentage (%) for categorical variables, means (m) and standard deviations (\(\pm\)) for continuous and normal distributed variables. Normal distribution was assessed by Shapiro-Wilk-Test.

**Results**

32 patients met the inclusion criteria of traumatic spinal injury which led to neurologic deficits without any other cerebral or skeletal injuries, who’s MRI scans revealed no pathologic findings. The patients were between 5 and 21 years old (mean: 14.5 years). There were 11 (34.4\% ) female and 21 (65.6\% ) male patients. 11 patients were admitted to the intensive care unit (ICU) (34.4\%) with a mean stay in the ICU of 1.3 days (1-3 days). The Glasgow Coma Scale (GCS) was 15 in all patients and the ASA physical status was I. The mean hospital stay was 2.75 days (1-13 days). No patient required surgical intervention. Most patients fell from height (43.8\%). 25\% of the patients were injured in a sporting accident, 18.8\% had a motor vehicle accident and 12.5\% of the patients got injured
Mechanism of injury

ASA physical status: 1.0 (1.0/1.0)

GCS: 15 (14/15)

Intensive care unit stay (in days): 1.3 (1-3)

Intensive care unit: relevant PTSD after “Real SCOWORA”.

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We found excellent clinical and functional results with a full neurologic recovery in all patients. However, a high number of 4/25 patients suffered from a relevant PTSD after “Real SCOWORA”.

Discussion

This is the largest series of patients evaluating mid- to long-term outcome after “Real SCIWORA” syndrome with complete absence of radiographic signs of trauma even in high-resolution MRI. We found excellent clinical and functional results with a full neurologic recovery in all patients. However, a high number of 4/25 patients suffered from a relevant PTSD after “Real SCOWORA”.

Table 1. Baseline Characteristics of the Study Population.

| SCIWORA patients | n = 32 |
|------------------|--------|
| Age (in years)   | 14.5 (5/21) |
| Gender           |         |
| Female           | 11 (34.4%) |
| Male             | 21 (65.6%) |
| Intensive care unit stay (in days) | 1.3 (1-3) |
| ASA physical status | 1.0 (1.0/1.0) |
| In hospital stay (in days) | 2.75 (1/13) |
| surgery yes      | 0 (0%) |
| no               | 32 (100%) |
| Mechanism of injury |       |
| motor vehicle accident | 6 (18.8%) |
| fall from height  | 14 (43.8%) |
| sporting accident | 8 (25.0%) |
| playing          | 4 (12.5%) |

Data show number of patients for categorical variables; ASA physical status (American Society of Anaesthesiologists Score); GCS (Glasgow Coma Scale).

The incidence of SCIWORA described in literature shows a wide range between 3.3% and 37%, depending on patients’ age and the use of MRI. The prevalence may be overestimated due to mis-classification and lack of MRI. The term “SCIWORA” is often inappropriately used as an umbrella term for clinical findings that are difficult to diagnose such as spinal cord concussion, brachial plexus, nerve root, or peripheral nerve injuries. Even though many articles deal with the topic of SCIWORA there is no consensus regarding the definition, demographics, etiology, treatment or prognosis.

SCIWORA is more commonly seen in the children and adolescents than in the older population, an observation we also made in our collective. Since SCIWORA often occurs as a result of hyperextension forces (e.g., cervical acceleration causing whiplash injuries in car accidents) or from a direct impact to the face, main causes of injury in literature for young children are road accidents, falls and child abuse. In adolescent years sporting accidents are getting more common. These observations are supported by our data as well as by reports about evolvement of injury mechanisms of children and adolescents in general.

Initial clinical assessment and anamnesis are more difficult with young patients with numerous factors (anxiety of patients and parents, lack of pediatric and adolescent clinical experience in emergency room, unconscious patient) complicating the first evaluation. Therefore initial management begins with spinal immobilization and clinical evaluation.

The initial neurological presentation is multifaceted, ranging from paresthesia, motor weakness, sensory loss, pathological reflexes, loss of rectal tone, incontinence and paralysis to...
Table 3. Mid-Term to Long-term Clinical Results After Real SCIWORA in the Study Population.

| SCIWORA Patients | n = 25 |
|-------------------|--------|
| Health related Quality of Life: | 98.48 |
| EQ-5D VAS | 0.957 |
| EQ-5D Index Value | |
| Oswemtry Disability Index | 6.04% (0-20%) |
| Posttraumatic stress disorder | 4/25 |
| (Breslau short screening scale) | |

Mid- to long-term outcome of 25 Real SCIWORA patients using the EQ-5D for the evaluation of health-related Quality of Life, the Oswestry Disability Index and the Breslau short screening scale for an evaluation of posttraumatic stress disorder.

The Advanced Trauma Life Support (ATLS) protocol is applied for traumatic spinal cord injuries (SCI), and therefore provides the basis of the treatment algorithm for patients with Real SCIWORA.

Our standardized protocol involves an interdisciplinary approach: The spine is immobilized, if it wasn’t already done in the field. The presence of a trauma surgeon, pediatrician and neurologist is advisable to detect subtle neurologic deficits that may be overlooked, especially in preverbal children. We provided neurologic clinical examination by a specialist in all patients with neurologic deficits and pediatric assistance for all patients under the age of 18. After clinical examination and stabilization of the patient radiologic evaluation was executed. The radiographic algorithm was based on trauma mechanism, age, clinical presentation, and hemodynamic stability. In patients with manifest neurologic symptoms and radiographs without pathologic findings, a MRI scan of the whole spine is mandatory. The duration of spinal immobilization is dependent on the recovery of neurologic symptoms and general clinical presentation but was generally recommended for 2-6 weeks in our population. In literature, there is no evidence for any advantage for longer immobilization in “Real SCIWORA.”

Regardless of neurological findings, there is no recommendation for any surgical intervention in “Real SCIWORA” patients, since any radiologic evidence of pathology is an indication for a more severe diagnosis than “Real-SCIWORA Syndrome.”

Pang et al., as well as several other authors, stated that initial neurological status is the only telltale sign of outcome. We reevaluated all patients 6 weeks after trauma and 25/32 after 6.9 years (1-14 years). None of the patients reported any relevant physical residue. In our cohort of only “Real SCIWORA” patients, we found that not the initial clinical impairment determines the outcome but the absence of pathologies in MRI. If the MRI shows any signs of injury there is a more accurate diagnosis than SCIWORA. In our cohort, a complete recovery was achieved after 3.9 days (1-13 days). This underlines the necessity of a clear diagnosing algorithm including a high-resolution MRI to confirm the diagnosis of “Real SCIWORA” per exclusionem of any other pathology.

To further characterize our patients we also collected data referring to the psychological impact of the trauma with neurologic symptoms depression, anxiety and PTSD is well described for SCI. The rate of PTSD for SCI is 7-44%. In our cohort 4 out of 25 patients reported symptoms of a PTSD even though there was no structural injury and all patients achieved complete recovery of neurologic symptoms. Therefore, we expanded our treatment algorithm and offer a psychologic co-treatment to “Real SCIWORA” patients. Surely, our study has some limitations. The main issue is the limited number of cases and the retrospective design. Even in a large level I trauma center, it was not possible to enroll more than 32 patients to the study. However, the strict inclusion criteria for the “Real SCIWORA” and exclusion of all peripheral reasons for neurologic impairments lead to this small number of cases. However, this is the first reasonable number of...
cases on mid-term outcome of “Real SCIWORA” in literature. Further studies are needed to substantiate the hypothesis of complete recovery for all cases of “Real SCIWORA” syndrome.

Conclusion
In this retrospective study we report on the largest series of only “Real SCIWORA” patients. With exclusion of any post-traumatic pathology in a high-resolution MRI, we observed full recovery in all patients. Therefore, we advocate for a diagnosis per exclusionem, a full spinal MRI is mandatory. Even though all patients recovered from neurologic symptoms, it is of utmost importance to evaluate each case to ensure detection of structural and potentially serious reasons for the neurologic condition. Psychological impact of neurologic disability and trauma are substantial and psychological support is advisable to prevent PTSD.

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Implications and Contributions
VF carried out data interpretation, performed literature research and drafted the manuscript. VA, KB, CS, JK and SL were involved in the design and correction work. FB was significantly involved in design, coordination and draft of the manuscript. All authors read and approved the final version of the manuscript.

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