Spinal Cord Injury Without Radiographic Abnormality (SCIWORA)

KEY WORDS: Pediatric patient population, SCIWORA, Spinal immobilization

DOI: 10.1227/NEU.0b013e3182770ebc
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RECOMMENDATIONS

Diagnosis:
Level III:
- Magnetic resonance imaging of the region of suspected neurological injury is recommended in a patient with spinal cord injury without radiographic abnormality (SCIWORA).
- Radiographic screening of the entire spinal column is recommended.
- Assessment of spinal stability in a SCIWORA patient is recommended with flexion-extension radiographs in the acute setting and at late follow-up, even in the presence of a magnetic resonance imaging negative for extraneural injury.
- Neither spinal angiography nor myelography is recommended in the evaluation of patients with SCIWORA.

Treatment:
Level III:
- External immobilization of the spinal segment of injury is recommended for up to 12 weeks.
- Early discontinuation of external immobilization is recommended for patients who become asymptomatic and in whom spinal stability is confirmed with flexion and extension radiographs.
- Avoidance of “high-risk” activities for up to 6 months following SCIWORA is recommended.

RATIONALE

Diagnosis
Pang and Wilberger1 defined the term spinal cord injury without radiographic abnormality (SCIWORA) in 1982 as “objective signs of myelopathy as a result of trauma” with no evidence of fracture or ligamentous instability on plain spine radiographs and tomography. The definition specifically excluded all magnetic resonance imaging (MRI) findings and any injuries from penetrating trauma, electric shock, and obstetrical complications and those associated with congenital spinal anomalies.2 Although many practitioners may consider this diagnostic terminology anachronistic in light of the current near-universal availability of MRI, pediatric neurosurgeons continue to refer to this predominantly pediatric phenomenon as SCIWORA.

In their original article, Pang and Wilberger cautioned, “If the early warning signs of transient symptoms could be recognized and promptly acted upon before the onset of neurological signs, the tragic fate of some of these children might be duly averted.” Hamilton and Myles,2 Osenbach and Menezes,3 and Pang and Wilberger1-3 have documented the delayed onset of SCIWORA in children as late as 4 days following injury. Therefore, a concern is whether a child with a normal neurological examination but with persisting subjective neurological symptoms referable to traumatic myelopathy should be assigned the diagnosis of SCIWORA and managed accordingly, despite the absence of “objective signs of myelopathy.”

Pang and Pollack4 have recommended obtaining a computed tomography scan focused at the neurological level of injury to exclude an occult...
fracture in a child with a neurological deficit referable to the spinal cord without abnormalities on plain radiographs of the spine. In addition, dynamic flexion and extension radiographs or fluoroscopy has been advocated to exclude pathological intersegmental motion consistent with ligamentous injury without fracture. If paraspinal muscle spasm, pain, or uncooperation prevents dynamic studies, they recommended external immobilization until the child can cooperatively flex and extend the spine for dynamic x-ray assessment. The finding of fracture, subluxation, or abnormal intersegmental motion at the level of neurological injury excludes SCIWORA as a diagnosis. In the initial report by Pang and Wilberger,1 1 of 24 children showed pathological motion on initial dynamic radiographs. By their own definition of SCIWORA, this 1 child would not be diagnosed with SCIWORA because the initial flexion and extension radiographs were abnormal. Although concern exists for the development of pathological intersegmental motion in children with SCIWORA following normal flexion and extension studies, there has been no documentation of such instability ever developing.

MRI findings in children with SCIWORA have spanned the spectrum from normal to complete cord disruption, along with evidence of ligamentous and disk injury in some.3–7 Possible roles for MRI of children with SCIWORA include identifying of signal change or intramedullary injury, excluding compressive lesions of the cord or roots or spinal ligamentous disruption that might warrant surgical intervention, guiding treatment regarding length of external immobilization, and/or determining when to allow patients to return to full activity.

Pang5 has also recommended somatosensory evoked potential (SSEP) screening of children with presumed SCIWORA. Possible roles for SSEPs in children with presumed SCIWORA include detecting subtle posterior column dysfunction when clinical findings are inconclusive, evaluating head-injured, comatose, or pharmacologically paralyzed children, distinguishing between intracranial, spinal, or peripheral nerve injuries, and/or providing a baseline MRI examination for comparison with subsequent evaluations.

Treatment

Because subluxation and/or malalignment are, by definition, absent in SCIWORA, the mainstay of treatment has been immobilization and avoidance of activity that may either lead to exacerbation of the present injury or increase the potential for recurrent injury. Medical management issues such as blood pressure support and pharmacological therapy are of concern to this population as well and have been addressed in other guidelines. (Of note, the often-cited prospective studies of pharmacological therapy in the treatment of acute spinal cord injuries did not include children < 13 years of age.5)

Pang and Pollack5 originally recommended 12 weeks of external immobilization to allow adequate time for the healing of the presumed ligamentous strain/injury and to prevent exacerbation of the myelopathy. Conversely, Bosch et al7 found no evidence that bracing prevents recurrent SCIWORA and therefore recommended bracing only on a case-by-case basis. It is unclear, however, what role immobilization plays in this population once dynamic radiographs have confirmed the absence of instability. The duration of and even the need for immobilization remain debatable given the current literature. If the incidence of delayed pathological intersegmental motion in children with SCIWORA who have been proven to have normal dynamic radiographs approaches zero, then the role of spinal immobilization for SCIWORA patients needs to be considered in light of the available literature. If (normal) physiological motion of the spinal column can potentiate spinal cord injury (SCIWORA) in these patients when there is no malalignment, subluxation, or lesion causing cord compression, then immobilization may be warranted in these patients.

Prognosis

SCIWORA has been shown to be associated with a high incidence of complete neurological injuries, particularly in children < 9 years of age. Hadley et al10 reported 4 complete injuries in 6 children < 10 years of age with SCIWORA. The regions of complete injury tend to be cervical and upper thoracic. Pang and Wilberger1 and Pang7 found the presenting neurological examination to relate strongly to outcome. Some data suggest that MRI abnormalities (or lack of abnormalities) of the cord may be more predictive of outcome than presenting neurological status.11-13 Because no child has been documented to develop spinal instability following the diagnosis of SCIWORA and has, by definition, normal flexion and extension radiographs, there has been little impetus to define predictors of instability. On the other hand, children have been documented to suffer recurrent SCIWORA,17,14 and predictors of a “high-risk” subgroup of children with SCIWORA for recurrent injury may exist. The guidelines author group of the Joint Section on Disorders of the Spine and Peripheral Nerves of the American Association of Neurological Surgeons and the Congress of Neurological Surgeons have previously produced a medical evidence-based guideline on this topic.15 The purpose of this updated review is to provide a contemporary analysis of the literature on the diagnosis and treatment of SCIWORA since that original publication.

SEARCH CRITERIA

A National Library of Medicine (PubMed) computerized literature search from 1966 to 2011 was undertaken using Medical Subject Headings in combination with “spinal cord injury”: “pediatric,” “spinal cord concussion,” “cervical cord neuropaxia,” and “SCIWORA.” Approximately 188 citations were acquired. Non-English language citations were deleted. Articles written in English were reviewed for those that identified children who incurred an SCIWORA. Those articles that described the clinical aspects and management of children with SCIWORA were used to generate these guidelines. Case reports were excluded from review. Of the 19 articles meeting selection criteria, none provided Class I or Class II medical evidence. All were case series representing Class III medical evidence. Summaries of these 19 articles are provided in Evidentiary Table format (Table 1).
TABLE 1. Evidentiary Table: Spinal Cord Injury Without Radiographic Abnormality*

<table>
<thead>
<tr>
<th>Reference</th>
<th>Description of Study</th>
<th>Evidence Class</th>
<th>Conclusions</th>
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<tr>
<td>Liao et al,23 Journal of Neurosurgery, 2005</td>
<td>Retrospective review of 9 SCIWORA patients &lt; 8 y of age correlating MRI findings with neurological outcomes</td>
<td>III</td>
<td>Prognosis correlated with MRI findings. Spinal cord transection and contusion were associated with severe, permanent deficits; normal MRI findings were associated with complete recovery.</td>
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<tr>
<td>Pang,7 Neurosurgery, 2004</td>
<td>Retrospective review of 95 pediatric SCIWORA cases (77 cervical, 18 thoracic; includes 55 reported in 1989); MRI and SSEP used to evaluate latter portion of the series</td>
<td>III</td>
<td>46 &quot;severe&quot; injuries, 49 &quot;mild&quot; injuries. Age &lt; 8 y associated with more severe injuries. SSEPs more sensitive than MRI in SCIWORA evaluation. Transient deficit or &quot;symptom only&quot; with normal MRI and SSEP treated in collar for 1-2 wk; all others with 12 wk of Guilford brace; no recurrent injuries.</td>
</tr>
<tr>
<td>Bosch et al,9 Spine, 2002</td>
<td>Retrospective review of 189 cases of pediatric SCIWORA over 35 y</td>
<td>III</td>
<td>Recurrent SCIWORA occurred in 21/189 cases and was not prevented by rigid bracing/immobilization after occult instability was properly ruled out.</td>
</tr>
<tr>
<td>Dare et al,11 Journal of Neurosurgery, 2002</td>
<td>Retrospective review of 20 pediatric SCIWORA cases evaluated with &quot;early&quot; MRI</td>
<td>III</td>
<td>MRI abnormalities seen only in those with complete neurological deficits (2/20). Conventional MRI sequences may lack sensitivity in cases of partial or transient deficits owing to SCIWORA.</td>
</tr>
<tr>
<td>Eleraky et al,16 Journal of Neurosurgery, 2000</td>
<td>Retrospective review of 102 children with cervical spinal injuries; young (0-9 y) compared with older children; MRI performed in 12/18 children with SCIWORA</td>
<td>III</td>
<td>SCIWORA in 18%. MRI findings did not alter management (external immobilization).</td>
</tr>
<tr>
<td>Turgut et al,26 European Spine Journal, 1996</td>
<td>Retrospective review of 11 of 82 children with spinal injuries with SCIWORA</td>
<td>III</td>
<td>SCIWORA represented 13% of spinal injuries in children.</td>
</tr>
<tr>
<td>Grabb and Pang,14 Neurosurgery 1994</td>
<td>Retrospective review of 7 children with SCIWORA who underwent MRI neurological status at presentation and follow-up was correlated to MRI findings</td>
<td>III</td>
<td>No compressive lesions found. Prognosis correlated with MRI findings. Hematomyelia involving &gt; 50% of cord diameter was associated with permanent severe deficits. Lesser degrees of hematomyelia and edema only were associated with incomplete recovery, and normal MRI predicted full recovery.</td>
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<tr>
<td>Davis et al,12 AJNR: American Journal of Neuroradiology, 1993</td>
<td>Retrospective review of 15 children with spinal cord injury who underwent MRI 12 h to 2 mo after injury, 7 with SCIWORA</td>
<td>III</td>
<td>MRI correlated with prognosis. Hemorrhagic cord contusions and cord &quot;infarction&quot; were associated with permanent deficits. No compressive lesions in SCIWORA cases. Normal MRI was associated with no myelopathy.</td>
</tr>
<tr>
<td>Hamilton and Myles,4 Journal of Neurosurgery, 1992</td>
<td>Retrospective review of 174 pediatric spinal injuries over 14-y period</td>
<td>III</td>
<td>SCIWORA represented 13% of spinal injuries. Of children 0 to 9 y of age with spinal injuries, 42% had SCIWORA, whereas of children 10 to 14 y of age, only 14% had SCIWORA.</td>
</tr>
<tr>
<td>Osenbach and Menezes,25 Neurosurgery 1992</td>
<td>Retrospective review of 34/179 children with spinal injuries with SCIWORA</td>
<td>III</td>
<td>SCIWORA represented 19% of spinal injuries in children. Younger children (&lt; 9 y) had higher incidence of SCIWORA.</td>
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<tr>
<td>Rathbone et al,27 Journal of Pediatric Orthopedics, 1992</td>
<td>Retrospective review of 12 children with presumed spinal cord concussion during athletics investigated for the presence of cervical stenosis</td>
<td>III</td>
<td>3 had a Torg ratio &lt; 0.8 and 4 had a canal anteroposterior diameter &lt; 13.4 mm. MRI was not used to evaluate for stenosis.</td>
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SCIENTIFIC FOUNDATION

One concern is whether the child with a normal neurological examination and either a history of transient neurological deficit (ie, paraparesis or quadriplegesis) or persisting subjective symptoms (ie, numbness or dysesthesias) would be a candidate for the diagnosis of SCIWORA. Pang and Wilberger described 13 of their 24 children to have a “latent” period from 30 minutes to 4 days (mean, 1.2 days) before the onset of objective sensorimotor deficits. All 13 of these children had transient subjective complaints at the time of their initial trauma that cleared within 1 hour before their subsequent neurological decline. Those who developed mild neurological deficits often improved to normal, and those who developed severe neurological deficits were often left with permanent neurological dysfunction. Hamilton and Myles, Osenbach and Menezes, and Pang and Pollack also reported a 22%, 23%, and 27% incidence, respectively, of delayed onset of myelopathy within their series of children with SCIWORA. Dickman et al, Eleraky and associates, and Hadley et al described no child having a latent period of neurological normalcy following injury. The observations of delayed deterioration by different investigators, however, raises the concern that any child presenting with a history of transient neurological...
deficit or symptoms following an appropriate mechanism of injury may be considered for the diagnosis of SCIWORA despite the absence of objective evidence of myelopathy on the initial neurological examination.

Pang and Wilberger reported 1 child of 24 whom they managed who demonstrated pathological intersegmental spinal motion on flexion and extension radiographs 1 week after injury, following resolution of that child’s neck pain and paraspinal muscle spasm. By definition, this child would not be considered to have had SCIWORA because the initial flexion and extension radiographs were abnormal. That child was treated successfully with external immobilization alone for 8 weeks. No child with SCIWORA has been documented in the literature to have had normal dynamic radiographs and then subsequently develop intersegmental instability.

In 1994, a series of 7 children with SCIWORA were demonstrated to have ligamentous, disk, and intramedullary abnormalities identified on MRI. Soft-tissue findings consisted of anterior longitudinal ligament disruption in association with a hyperextension injury, posterior longitudinal ligament disruption, and a non-compressive C2-3 disk herniation associated with lateral flexion and 1 case of C6-7 disk abnormality associated with hyperflexion. Intramedullary findings reported included cord transection and rostral stump hemorrhage, severe hematomyelia, a minor intramedullary hemorrhage, and edema without hemorrhage. Davis et al described 7 children with SCIWORA who were imaged with MRI. They described no abnormalities of muscles, ligaments, or disks but correlated the presence of intramedullary hemorrhage or cord “infarction” with permanent neurological deficit. The lack of intramedullary findings correlated with a normal neurological outcome. Dickman et al commented on 7 children with SCIWORA who were imaged with MRI. Five of the 7 studies revealed no abnormality, and 2 studies documented intramedullary signal changes. Osenbach and Menezes commented in their series of childhood SCIWORA that MRI and computed tomography–myelography performed on their patients did not demonstrate a single compressive lesion. In addition, they performed spinal arteriograms in 4 of 5 children with thoracic SCIWORA and found no angiographic abnormalities. Rossitch and Oakes performed myelograms on neonates and found no abnormalities that changed their management. Hadley et al obtained MRIs before 1988 on 5 children with SCIWORA and identified no abnormalities. Date et al reviewed early MRI studies on 20 consecutive pediatric SCIWORA patients from 1992 to 1999, reporting abnormalities only in those with complete neurologic deficits. These reports and their results need to be viewed in the context of the technology and image quality available at the time of investigation.

SSEP evaluations of pediatric SCIWORA have been reported by Pang. In his series of 50 pediatric SCIWORA patients evaluated with both MRI and SSEPs, the SSEPs demonstrated abnormalities more frequently than MRI for both permanent and transient deficits. Pang is the only investigator to report any alteration in the clinical management of pediatric SCIWORA based on MRI/SSEP findings. No child with MRI-documented ligamentous injury and SCIWORA has developed spinal instability, early or delayed. There has been no correlation between the ligamentous findings on MRI in SCIWORA patients and subsequent spinal instability to date. The appearance of the spinal cord on MRI does provide prognostic information regarding ultimate neurological outcome.

Hadley et al noted a 16% incidence of multiple noncontiguous injuries of the spine or spinal cord in children with any type of spinal column or spinal cord injury. Ruge et al had a similar incidence (17%) of multiple levels of spinal injury in children. Although neither of these 2 studies dealt with an isolated population of children with SCIWORA, they provide consistent observations that SCIWORA following admission or discharge. None of these 3 reports described the length of time children with SCIWORA were immobilized. Bosch et al reviewed 189 cases of pediatric SCIWORA over 35 years, reporting no instances of neurologic deterioration and no benefit to bracing because it did not appear to reduce the frequency of recurrent SCIWORA. It has not been routine among treating physicians to prescribe 12 weeks of immobilization for children with SCIWORA. Although the single report by Pollack et al describes recurrent SCIWORA within 12 weeks of the original injury, this has not been validated by other observations. Because MRI evaluation was not available...
for those with recurrent injury, it is not known whether certain MRI characteristics (eg, ligamentous disruption) could predict an “at-risk” group for recurrent SCIWORA.

Additional case series of mild, transient SCIWORA have been reported using different terminology, namely “spinal cord concussion” or “cervical cord neurapraxia.”19-22 The diagnostic criteria for these entities fall within the definition of SCIWORA and further require that the symptoms/deficits resolve within 48 to 72 hours. Most of the patients in these reports are adolescents and young adults injured during athletic activities, especially American tackle football. Many of these series report an association between radiographically documented spinal stenosis and recurrent episodes20-22; however, Pang asserts that “congenital cervical stenosis and resultant [spinal cord injury], such as in young athletes, most definitely should be excluded from the SCIWORA umbrella” (D. Pang, MD, personal communication, November 2010).7 Return-to-play recommendations proposed in these reports are arbitrary, controversial, and beyond the scope of these guidelines.

While Pang and Wilberger1 reported that in their series neurological outcome correlated with the presenting neurological status, the MRI appearance of the spinal cord has been shown to be predictive of neurological outcome in children with SCIWORA.7,11-13,23 Absence of signal change within the cord is associated with an excellent outcome. Signal change consistent with edema or microhemorrhages, but not frank hematomyelia, is associated with significant improvement of neurological function over time. The presence of frank hematomyelia or cord disruption is associated with a severe, permanent neurological injury.11-13 The correlation of neurological outcome with spinal cord MRI findings in SCIWORA remains consistent with the findings in much larger numbers of patients with spinal cord injury (non-SCIWORA) who have been studied with MRI.4,23 Recognized prognostic factors related to SCIWORA are summarized in Table 2.

SUMMARY

SCIWORA is a widely recognized form of spinal cord injury, occurring almost exclusively in children, and is characterized by the absence of any radiographically evident fracture, dislocation, or malalignment. Children presenting with a history of transient neurological signs or symptoms referable to the spinal cord after a traumatic event, despite the absence of objective neurological deficits with normal radiographs, may develop SCIWORA in a delayed fashion.

No child with SCIWORA has developed pathological intersegmental motion with instability when early flexion and extension radiographs have been normal.

MRI has not identified any abnormal findings in a child with SCIWORA when the management scheme would be changed by the results of the MRI. Similarly, no child with SCIWORA in whom a subsequent MRI has documented ligamentous injury has developed evidence of spinal instability.

Treatment consisting of cervicothoracic bracing for patients with cervical-level SCIWORA for 12 weeks and avoidance of activities that encourage flexion and extension of the neck for an additional 12 weeks has not been associated with recurrent injury. Patients with normal MRI and SSEP findings following transient deficits or “symptoms only” may be managed with a cervical collar for 1 to 2 weeks.

The spinal cord findings on MRI provide prognostic information regarding long-term neurological outcome in patients with SCIWORA. Myelography and angiography have no defined role in the evaluation of children with SCIWORA.

KEY ISSUES FOR FUTURE INVESTIGATION

The treatment end points of spinal immobilization and activity restriction for patients with SCIWORA have been arbitrarily chosen. MRI may be helpful to guide the duration of immobilization and activity restriction for that child. For example, the absence of ligamentous injury by MRI may indicate that there is no need for external immobilization or activity restriction. It has been observed that SCIWORA can recur despite the absence of demonstrable spinal instability and may not be prevented by bracing. A study that obtained MRI on all children with SCIWORA and followed up their clinical status longitudinally may highlight the utility of MRI in the management of children who go on to develop recurrent SCIWORA.

The literature provides little guidance as to the likelihood for subsequent catastrophic injury in children presenting with SCIWORA of any severity who are found to have a preexisting spinal or neurological abnormality such as congenital cervical stenosis or a Chiari malformation.19,21,24 Longitudinal clinical follow-up of SCIWORA patients of this type may provide information to appropriately counsel these children.

<table>
<thead>
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<th>TABLE 2. Recognized Prognostic Factors in SCIWORAa</th>
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<td>Spinal cord injury without radiographic abnormality presenting with a complete or severe neurologic deficit has a poor prognosis.</td>
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<tr>
<td>Mild, partial spinal cord injury without radiographic abnormality (spinal cord concussion) usually recovers to normal function.</td>
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<tr>
<td>Recurrent spinal cord injury without radiographic abnormality appears to be rare and usually occurs within 2 weeks of presentation.</td>
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<tr>
<td>Magnetic resonance imaging evidence of spinal cord disruption or major hemorrhage is strongly associated with complete or severe neurologic deficit and carries a poor prognosis.</td>
</tr>
<tr>
<td>A normal magnetic resonance image of the region of neurological injury is associated with a favorable prognosis.</td>
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</table>

aSCIWORA, spinal cord injury without radiographic abnormality.
There are no data to elucidate the role of age in the success or failure of various treatments for this condition. This could be undertaken in a longitudinal study of a patient population of reasonable size. Serious attempts to address the topics above cannot be forthcoming from a single institution or investigator because of the relatively low numbers of children who sustain SCIWORA annually.25,26 A multiple-institution protocol-directed study of SCIWORA patients may provide answers to some of the questions that accompany this unique spinal cord injury subtype.

Disclosure

The authors have no personal financial or institutional interest in any of the drugs, materials, or devices described in this article.

REFERENCES