

SPINAL CORD INJURY WITHOUT RADIOGRAPHIC ABNORMALITY (SCIWORA)

RECOMMENDATIONS

Diagnosis:

Standards: There is insufficient evidence to support diagnostic standards.

Guidelines: There is insufficient evidence to support diagnostic guidelines.

Options:

- Plain spinal radiographs of the region of injury and CT scan with attention to the suspected level of neurological injury to exclude occult fractures are recommended.
- MR of the region of suspected neurological injury may provide useful diagnostic information.
- Plain radiographs of the entire spinal column may be considered.
- Neither spinal angiography nor myelography is recommended in the evaluation of patients with SCIWORA.

Treatment:

Standards: There is insufficient evidence to support treatment standards.

Guidelines: There is insufficient evidence to support treatment guidelines.

Options:

- External immobilization is recommended until spinal stability is confirmed flexion and extension radiographs.
- External immobilization of the spinal segment of injury for up to 12 weeks may be considered.
- Avoidance of “high-risk” activities for up to six months following SCIWORA may be considered.

Prognosis:

Standards: There is insufficient evidence to support prognostic standards.

Guidelines: There is insufficient evidence to support prognostic guidelines.

Options: MRI of the region of neurological injury may provide useful prognostic information about neurological outcome following SCIWORA.

RATIONALEDiagnosis:

Pang and Wilberger defined the term SCIWORA (Spinal Cord Injury Without Radiographic Abnormality) 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.(11) In their original manuscript they cautioned, “that 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”.(11) Hamilton and Myles, Osenbach and Menezes, and Pang and Wilberger, (8,9,11) have documented the delayed onset of SCIWORA in children as late as four days following injury. Therefore, a concern is whether a child with a normal neurological examination, but with a history of transient neurological symptoms or 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 Pollack have recommended obtaining a CT 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.(12) In addition, dynamic flexion and extension radiographs or fluoroscopy have been advocated to exclude pathological

intersegmental motion consistent with ligamentous injury without fracture. If paraspinous muscle spasm, pain, or uncooperation prevents dynamic studies, they recommended external immobilization until the child can 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, one of 24 children showed pathological motion on initial dynamic radiographs.(11) By their own definition of SCIWORA, this one child would not be diagnosed with SCIWORA because the initial flexion and extension radiographs were abnormal. While concern exists for the development of pathological intersegmental motion in children with SCIWORA following normal flexion and extension studies, there has not been documentation of such instability ever developing.

Magnetic resonance (MR) imaging findings in children with SCIWORA have spanned the spectrum from normal to complete cord disruption, along with evidence of ligamentous and disc injury in some.(3,6) Possible roles for MR of children with SCIWORA include: 1) exclude compressive lesions of the cord or roots or ligamentous disruption that might warrant surgical intervention, 2) guide treatment regarding length of external immobilization, and/or 3) determine when to allow patients to return to full activity.

Treatment:

Because there exists no subluxation or malalignment in SCIWORA the mainstay of treatment has been immobilization and avoidance of activity that may lead to either 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 injury did not include children younger than 13 years of age). (1)

Pang and Pollack have 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.(12) It is unclear however, what role immobilization plays in this population once dynamic radiographs have displayed no instability. The length 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 physiological motion (normal) 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 is warranted in these patients.

Prognosis:

SCIWORA has been shown to be associated with a high incidence of complete neurological injuries, particularly in children less than nine years of age. Hadley, et al, reported four complete injuries in six children less than ten-years-old with SCIWORA.(7) The regions of complete injury tend to be cervical and upper thoracic. Pang found the presenting neurological examination to relate strongly to outcome.(11) There is some data to suggest that MR abnormalities (or lack of abnormalities) of the cord may be more predictive of outcome than presenting neurological status.(3,6) 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 (13), and predictors of a “high-risk” sub-group of children with SCIWORA for recurrent injury may exist.

SEARCH CRITERIA

A National Library of Medicine computerized literature search from 1966 to 2001 was undertaken using Medical Subject Headings in combination with “spinal cord injury”: “pediatric” and “SCIWORA.” Approximately 145 citations were acquired. Non-English language citations were deleted. Articles written in English were reviewed for those that identified children that incurred a spinal cord injury without radiographic abnormality (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 fifteen articles meeting selection criteria, none were Class I or Class II studies. All were case series representing Class III data. They are summarized in Evidentiary Table format.

SCIENTIFIC FOUNDATION

One concern is whether the child with a normal neurological examination and either a history of transient neurological deficit (i.e. paraparesis or quadriparesis), or persisting subjective symptoms (i.e. 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 four days (mean 1.2 days) before the onset of objective sensorimotor deficits.(11) All 13 of these children had transient subjective complaints at the time of their initial trauma that cleared within one hour prior to their neurological decline. Those who developed mild neurological deficits often improved to normal, while those that developed severe neurological deficits were

often left with permanent neurological dysfunction. Hamilton and Myles, Osenbach and Menezes, and Pang and Pollack as well reported a 22, 23, and 27% incidence of delayed onset of myelopathy within their series of children with SCIWORA, respectively.(8,9,12) Dickman, et al, Eleraky and associates, and Hadley, et al, described no child having a latent period of neurological normalcy following injury.(4,5,7) 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 upon initial neurological examination.

Pang and Wilberger had one child out of 24 demonstrate what was considered to be pathological intersegmental motion on flexion and extension radiographs one week after injury, following resolution of neck pain and paraspinous muscle spasm.(11) By definition this child would not be considered to have had SCIWORA, because the initial flexion and extension radiographs were abnormal. This child was treated successfully with external immobilization alone for eight 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 seven children with SCIWORA were demonstrated to have ligamentous, disc, and intramedullary abnormalities identified on MRI imaging.(6) Soft tissue findings consisted of anterior longitudinal ligament disruption in association with a hyperextension injury, posterior longitudinal ligament disruption and a noncompressive C2-3 disc herniation associated with lateral flexion, and one case of C6-7 disc 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 seven children with SCIWORA who were imaged with MRI.(3) They described no abnormalities of muscles, ligaments, or discs, but did correlate 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 seven children with SCIWORA who were imaged with MR. Five of the seven studies revealed no abnormality and two studies documented intramedullary signal changes.(4) Osenbach and Menezes commented in their series of childhood SCIWORA that MRI and CT-myelography performed on their patients did not demonstrate a single compressive lesion.(9) In addition, they performed spinal arteriograms in four of five children with thoracic SCIWORA and found no angiographic abnormalities. Rossitch and Oakes performed myelograms on neonates with SCIWORA from birth injury and found no abnormalities that changed their management.(16) Hadley et al, obtained MR imaging prior to 1988 on five children with SCIWORA and identified no abnormalities.(7) These results need to be viewed in the context of the technology available at the time of study.

There has been no report of any situation in which the care of a child with SCIWORA has been altered by the results of MR and/or myelography imaging studies. No child with MR 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 MR does provide prognostic information regarding ultimate neurological outcome.

Hadley, et al, noted a 16% incidence of multiple non-contiguous injuries of the spine or spinal cord in children with any type of spinal column or spinal cord injury.(7) Ruge, et al, had a similar incidence (17%) of multiple levels of spinal injury in children.(17) While neither of

these two studies dealt with an isolated population of children with SCIWORA, they do provide consistent observations that one in six children with spinal trauma will have multiple levels of injury. Pang and Wilberger reported one of 24 children with a second level injury (L2 Chance fracture) who had a T6 neural injury (SCIWORA), but they did not obtain complete spine radiographs on every child.(11) Because of these observations one should consider radiographs of the entire spinal column when any traumatic spinal injury, SCIWORA or otherwise, is identified in a child.

In the initial series of children with SCIWORA reported by Pang and Wilberger, treatment routinely consisted of four weeks of external immobilization with a “cervical collar” for cervical injuries.(11) In cases of thoracic injury, if repeat plain radiographs showed no abnormality following one week of bed rest, the child was mobilized without a brace. In a later report in 1989, Pang and Pollack recommended 12 weeks of external immobilization for SCIWORA patients to allow for healing of the presumed ligamentous strain/injury, and to prevent exacerbation of the myelopathy.(12) They also advocated external immobilization for this time frame to prevent recurrent injury during the healing phase. They reported seven children who sustained recurrent SCIWORA of greater severity with lesser degrees of force when external immobilization was removed before 12 weeks time, or they were allowed to participate in activities against physician instructions within six months of the initial injury. For these reasons, they recommend 12 weeks of external immobilization and 12 additional weeks of activity restriction following SCIWORA.

Dickman et al, Eleraky et al, and Hadley and colleagues reported no neurological deterioration in any patient with SCIWORA following admission or discharge.(4,5,7) None of these three reports described the length of time children with SCIWORA were immobilized. It

has not been routine among treating physicians to prescribe 12 weeks of immobilization for children with SCIWORA.(2) While the single report by Pollack and Pang describes recurrent SCIWORA within 12 weeks of the original injury, this has not been validated by other observations.(13) Because MR evaluation was not available for those with recurrent injury, it is not known whether certain MR characteristics (i.e. ligamentous disruption) could predict an “at risk” group for recurrent SCIWORA.

While Pang and Wilberger reported that in their series, neurological outcome correlated with the presenting neurological status, the MR appearance of the spinal cord has been shown to be predictive of neurological outcome in children with SCIWORA.(3,6,11) Absence of signal change within the cord is associated with an excellent outcome. Signal change consistent with edema or micro-hemorrhages, 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.(3,6) The correlation of neurological outcome with spinal cord MR findings in SCIWORA remain consistent with the findings in much larger numbers of patients with spinal cord injury (non-SCIWORA) who have been studied with MR.(15)

SUMMARY

Children presenting with a history of transient neurological signs or symptoms referable to traumatic myelopathy despite the absence of objective evidence of myelopathy and normal radiographs may develop SCIWORA in a delayed fashion.

No child with SCIWORA has developed pathological intersegmental motion with instability after demonstrating normal flexion and extension radiographs.

MR has not identified any lesion in a child with SCIWORA where the management scheme would be changed by the results of the MR. Similarly, no child with MRI documented ligamentous injury and SCIWORA has developed evidence of spinal instability.

Hard collar immobilization 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.

The spinal cord findings on MRI imaging 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 endpoints of spinal immobilization and activity restriction for patients with SCIWORA have been arbitrarily chosen. MR may be helpful to guide the length of time a child is immobilized and activities restricted. The absence of ligamentous injury by MR may indicate that there is no need for external immobilization or activity restriction. It has been observed that recurrent SCIWORA can occur despite no evidence of spinal instability. An investigation that obtained MR imaging on all children with SCIWORA and followed their clinical status longitudinally, may highlight the utility of MR 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 pre-existing spinal or neurological abnormality such as congenital cervical stenosis or a Chiari

malformation.(14) Longitudinal clinical follow-up of SCIWORA patients of this type may provide information to appropriately counsel these children.

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 (10,18). A multiple-institution protocol directed study of SCIWORA patients may provide answers to some of the questions which accompany this unique spinal cord injury subtype.

EVIDENTIARY TABLES

| Authors & Year | Description of Study | Class of Data | Results |
|--|--|---------------|---|
| Eleraky MA et al, <i>J Neurosurg (Spine)</i> 2000 | Retrospective review of 102 children with cervical spinal injuries. Young (0-9 years) compared to older children. MR performed in 12 of 18 children with SCIWORA. | III | SCIWORA in 18%. MR findings did not alter management (external immobilization). |
| Turgut M et al, <i>Eur Spine J</i> , 1996 | Retrospective review of 11 of 82 children with spinal injuries with SCIWORA | III | SCIWORA represented 13% of spinal injuries in children. |
| Grabb PA & Pang D, <i>Neurosurgery</i> 1994 | Retrospective review of seven children with SCIWORA underwent MR. Neurological status at presentation and follow-up was correlated to MR findings. | III | No compressive lesions found. Prognosis correlated with MR findings. Hematomyelia involving greater than 50% of cord diameter was associated with permanent severe deficits. Lesser degrees of hematomyelia and edema only were associated with incomplete recovery, and normal MR predicted full recovery. |
| Davis PC et al, <i>AJNR</i> , 1993 | Retrospective review of 15 children with spinal cord injury underwent MR 12 hours to two months after injury. seven with SCIWORA | III | MR correlated with prognosis. Hemorrhagic cord contusions and cord “infarction” were associated with permanent deficits. No compressive lesions in SCIWORA cases. Normal MR was associated with no myelopathy |
| Hamilton MG & Myles ST, <i>J Neurosurg</i> , 1992 | Retrospective review of 174 pediatric spinal injuries over 14 year period | III | SCIWORA represented 13% of spinal injuries. Of children aged 0-9 years with spinal injuries 42% had SCIWORA, whereas children aged 10-14 years only 14% had SCIWORA. |
| Osenbach RK & Menezes AH, <i>Neurosurgery</i> 1992 | Retrospective review of 34 of 179 children with spinal injuries with SCIWORA | III | SCIWORA represented 19% of spinal injuries in children. Younger children (<9 years-old) had higher incidence of SCIWORA. |
| Rathbone D et al, <i>J Ped Orthop</i> , 1992 | Retrospective review of 12 children with presumed spinal cord concussion during athletics was investigated for the presence of cervical stenosis. | III | 3 had a Torg ratio < 0.8 and four had a canal AP diameter <13.4 mm. MRI was not used to evaluate for stenosis. |
| Rossitch E & Oakes WJ, <i>Pediatr Neurosurg</i> 1992 | Retrospective review of five neonates with perinatal spinal cord injury. four of the five had no abnormality on static spinal radiographs. No flexion/extension views reported. Myelograms were unrevealing. | III | Perinatal spinal cord injury often has normal radiographs. The neonates are often initially misdiagnosed. Respiratory insufficiency and hypotonia are common signs. |
| Dickman CA et al, <i>J Spinal Disorders</i> , 1991 | Retrospective review of 26 children with SCIWORA over 19-year period. Clinical and epidemiological features were analyzed. | III | SCIWORA 16% of spinal injuries in children. MVA was most common mechanism. Seven children had MR. five were normal studies, two showed cord signal abnormalities. Younger children tended to have more severe injuries. |

| Authors & Year | Description of Study | Class of Data | Results |
|--|--|----------------------|--|
| Osenbach RK & Menezes AH, <i>Pediatr Neurosci</i> , 1989 | Retrospective review of 31 children with SCIWORA | III | 26 cervical and five thoracic injuries. Complete cord injury in 12. Delayed onset of deficits in 7. No surgical lesions found by MR or CT-myelography. Spinal angiograms done in four thoracic cases were normal. No delayed instability at follow-up. |
| Pang D & Pollack IF, <i>J Trauma</i> , 1989 | Retrospective review of 55 children with SCIWORA (43 cervical, 12 thoracic). Clinical profiles reported to illustrate syndrome | III | 22 "severe" injuries 33 "mild" injuries <8 yo associated with more severe injuries 8 cases of recurrent injury from three days to ten weeks after initial injury No recurrent injuries with 12 weeks of Guilford Brace |
| Hadley MN et al, <i>J Neurosurg</i> , 1988 | Retrospective review of 122 children with spinal injuries. Young (0-9 years) compared to older children. | III | 17% with SCIWORA. Higher incidence of SCIWORA in 0-9 year-olds versus 10-16 year-olds. five studied with MRI, no abnormalities detected. |
| Pollack IF et al, <i>J Neurosurg</i> , 1988 | Retrospective review of eight children with recurrent SCIWORA compared to 12 children treated with longer immobilization | III | Recurrent SCIWORA occurred from three days to ten weeks after initial injury. Recurrent injuries were more severe. No recurrent injuries with 12 weeks of Guilford Brace. |
| Ruge JR et al, <i>J Neurosurg</i> , 1988 | Retrospective review comparing 0-3 year-olds to 4-12 year-olds with spinal injury | III | N=47, 21% with SCIWORA |
| Pang D & Wilberger JE, <i>J Neurosurg</i> , 1982 | Retrospective review of 24 children with SCIWORA | III | One child with instability on flex/ext at one week |

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