Epidemiology and management of spinal cord injury in children and adolescents

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ABSTRACT

Pediatric spinal cord injury (SCI) is a rare condition with significant long-term consequences affecting both children's and their caregivers' lives. Leading causes of pediatric SCI are motor vehicle accidents, falls and sport accidents. Patient assessment and management should be based in pediatric physiology and be adjusted to age. The maturity process brings up many unique challenges and complications such as scoliosis and hip dysplasia that have been reported to be common problems after sustaining a SCI in young age. Decreased mobility, autonomic dysreflexia, neurogenic bladder and neurogenic bowel have also all to be managed throughout the child's growing and maturity process. Rehabilitation and management of pediatric SCI patients aims in facilitating independence to meet peer lifestyle and successful transfer into adulthood. The present study is a literature review aiming to summarize the current knowledge on the rehabilitation and management of the secondary complications of pediatric onset SCI. A literature research was performed using the PubMed and Google Scholar online data bases and the following key words: "spinal cord injury," "pediatric", "children", "epidemiology", "management" and "rehabilitation". Following the PRISMA guidelines, 27 articles were finally included in this review. Caring for children or adolescents with SCI presents many unique secondary health complications and should be based on normal life milestones.

Key Words: spinal cord injury; pediatric; epidemiology; management

Introduction

Spinal cord injury (SCI) is damage to the spinal cord that leads to a medically complex condition with life-changing impact on the affected individuals. [1,2] As in SCI in adults, the degree of impaired mobility and secondary health complications in pediatric population SCI depends on the severity and the location of the injury in the spinal cord. However, children through all life stages until the completion of adolescence and the transition into adulthood have different, and sometimes unique, health complications from those occurring in adults. Age at injury, skeletal maturity and the ongoing dynamic developmental physiology throughout childhood and adolescence are factors to be considered in the rehabilitation approaches of SCI pediatric population. [1, 2, 3, 4] Life expectancy after SCI continuous to increase; however, patients that sustain SCI in an age younger than 16 years are

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noted to have reduced life expectancies compared to older ones. This fact is supposed to be the result of longer exposure to SCI complications and maximizes the need of long-term patient monitoring to prevent such complications. [1, 3]

Child's typical developmental motor milestones, communication and socialization skills are the main domains to consider adjusting the assessment and caring to patient's age. Depending on age at injury, certain skills may have never been achieved previously and they are taught for first time. Furthermore, undeveloped communication skills may present as barriers in the assessment and management of pediatric SCI patients and a more understandable language or the help of the caregivers may be required. [1] For example, assessment with the International Standards for Neurological Classification of Spinal Cord Injury (ISNCSCI) is recommended for children over 6 years old based on several largescale pediatric psychometric studies. [4] Children younger than 6 years old are not able to fully understand the directions given for the sensory examination and distinguish between normal and reduced sensation. [3,5]

The present study is a literature review aiming to summarize the current knowledge on the rehabilitation and management of the secondary complications of pediatric onset SCI. Following the PRISMA guidelines, 27 articles were finally included in this review as it is seen in Table 1. [8] Starting from year 2010, a literature research was performed by using the PubMed and Google Scholar online data bases and the following key words: "spinal cord injury," "pediatric", "children", "epidemiology", "management" and "rehabilitation" with Boolean operators. Animal studies, case reports, case series involving less than 10 participants, and studies involving participants with congenital spinal injury (e.g. spina bifida) or cerebral palsy were also excluded. Original and review articles written in English concerning SCI in children and adolescents (less than 18 years) were reviewed for epidemiological data, rehabilitation and secondary complications. Studies in adults were included if they had separate reporting for participants less than 18 years old. Studies in adults with pediatric-onset SCI were also excluded.

Discussion

Epidemiology

It is estimated that every year, around the world, between 250.000 and 500.000 people sustain a SCI. [6] It is most likely to occur in young individuals between the ages of 16 and 30 years (47% of all injuries) and the elderly (\geq 60 years old). Estimates reflect a consistent trend toward older age at time of injury. [6, 9] Children younger than 15 years old represent less than 10% of SCI cases. [3, 11] In more detail, incidence of pediatric SCI has been reported to be 5.27 per million per year between 1988 and 2014 in Spain (internal registry of the Spinal Cord Injury Unit) [12], 17.5 per million per year between 2007 and 2010 in the United States (Nationwide Emergency Department Sample) [9], and 5.99 per 100,000 per year between 1998 and 2008 in Taiwan (National Health Insurance Research Database) [13]. Studies with cut of age at 15 years old reported incidence ranging from 0 to 3.1 per million between 2000 and 2015 in Ireland (National Rehabilitation Hospital) [14] and 3.8 per million per year between 2000 and 2010 in Australia (Royal Children's Hospital). [15]

Pediatric patients are associated with higher risk of cervical SCI (approximately 80%) and SCI without radiological abnormality (SCIWORA) because of the underdeveloped anatomic characteristics of their spine. [3, 6, 16] The most common cause of pediatric SCI are motor vehicle accidents (MVA), while other common causes are falls and sports injuries. [9, 12, 13, 14, 15, 17, 18] Young children also sustain SCI through medical or surgical causes. [10,14] Sports accidents that result in SCI are more frequent in children older than 10 years old, with the higher rates being reported in the 10-13 years old age group. [17, 18] Other reported causes in adolescents include violence, firearm and self-inflicted injuries. [9, 15]

Males and females under 5 years of age seem to have the same risk sustaining SCI, while as the age at the time of injury increases males have a higher risk. [9, 10] This is probably due to males' increased risk behaviors especially during adolescence and young adulthood such as motorcycle riding, diving and involvement in violence/gunshots. [12, 13] The incidence of pediatric SCI has shown a downward trend in recent years. [9, 12, 14] This is probably related to road safety strategies

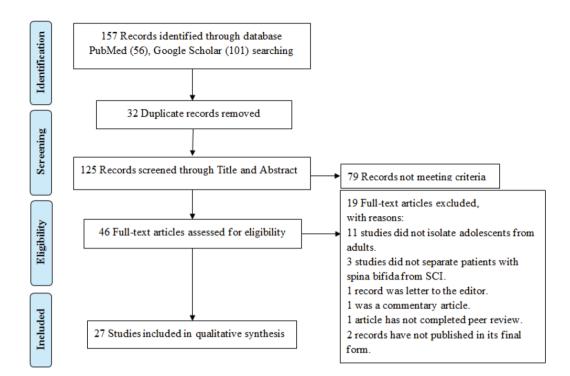


Table 1. Flow chart

having a positive impact in traffic incidents. [12, 14] Mortality rates are higher in children with SCI associated with cervical injury and MVA. [6, 9]

SCIWORA

Forces acting on the head and neck can result in great stretching of the spinal ligaments without resultant fracture but with injury to the spinal cord known as SCI without radiographic abnormalities (SCIWORA), which is more common in children under 8 years old. [3, 7] A multicenter retrospective study conducted in France between January 1988 and June 2017 included 37 patients (30 patients with SCIWORA, 2 patients with severe cranial trauma and 5 patients with obstetric trauma). Among SCIWORA patients, 50% of injuries were caused in MVA and 36.6% in sports accidents. Younger aged children sustained SCIWORA during MVA and children older than 7 years and adolescents during sport accidents. [16] Similarly, MVA and sports accidents were the main causes of injury among 297 SCIWORA patients included in the 2012 dataset of Healthcare Utilization Project KID (HCUP-

KID). In more detail, it was reported that children with SCIWORA under the age of 3 years usually were involved in MVA (38%) and falls (23%), while the main cause for SCIWORA in adolescents were sports accidents (57%) and MVA (19%). In all age groups, cervical spine was the most common location of injury. Upper cervical spine injuries, located between C1 and C4, were most common in the youngest patients (aged 0-3 years) while SCIWORA in adolescents often occurred by injury in the lower cervical and thoracic spine. [19]

Conservative and surgical treatment of pediatric SCI

The majority of pediatric cases with SCI are managed conservatively and surgical intervention is chosen if needed depending on the type of spine trauma and the age of the children. [6, 11] There is low evidence for using instrumentation in pediatric SCI, but there is literature that supports their safe and effective use in pediatric spinal deformities. Based on that literature, surgical stabilization is suggested in unstable pediatric trauma to protect neural elements [11] Although there are pharmacological treatments for adults who

sustained SCI, there is no evidence regarding the use of these treatments in children with SCI. [20, 21] However, studies investigating possible complications after high-dose steroid treatment in children with SCI reported no significantly high rates of complications. [20, 22]

Neurological Recovery

Neurological and functional outcomes after SCI in pediatric patients seem to be better when compared to the outcomes in adults. Although existing evidence is not significant enough to prove that, some studies reported good neurological recovery after SCI, with incomplete lesions having the best prognosis and complete lesions improving over time. [21] In a retrospective, multi-center case control study, each one of the 32 adolescents with SCI was matched to 2 adults according to neurological level of injury, initial AIS grade and total Spinal Cord Independence Measure (tSCIM). There were no statistical differences in respect to AIS conversion, but there were statistically significant higher final tSCIM scores in the adolescent group indicating higher functional improvement than adults. [2]

Ambulation - Gait training

In pediatric population with SCI mobility varies depending on age and impairment. The means used for mobility are evaluated and revised as needed during child's growth. The type of power or manual wheelchair needed depends on child's age, level of injury and functional status, environment, and child's and family's preferences. [24] Children about 1 year old should be encouraged to use a power or manual wheelchair to facilitate independence and limit the use of a stroller. Scooters are not considered an appropriate choice for most children and adolescents with SCI. [24] Among 131 children with SCI under 5 years of age using a wheelchair 98% were independent in propulsion, 3% exclusively used power wheelchairs, 85% exclusively used manual wheelchairs and 12% used both power and manual wheelchairs. Manual wheelchair use was initiated at a median age of 3 years and 5 months and power wheelchair use was initiated at a median age of 2 years and 11 months. [10]

Orthotics, assistive devices and locomotor training (LT) is an option for household and community ambu-

lation. [24] In a recent study, the walking ability of 48 ambulant children with SCI was measured using the WISCI score (ability of a person to walk 10m after a SCI from the most to the least severe impairment). The authors reported that 60.41% of patients had WISCI score 20, which means that more than half of children aged between 2-18 years were walking with no help (device, brace, physical assistance), and 14.58% had WISCI score 12, which means that patients walked with two crutches and two ankle foot orthosis. [18]

Recovery of walking is dependent on the age at the time of injury, the level of injury, and the completeness of neurologic impairment. [23] Younger children have better prognosis for recovery. Indeed, children injured at the age of 5 years or younger are approximately seven times more likely to walk than older ones. [23] Individuals with incomplete lesions are more likely to recover ambulation compared to those with complete lesions. [23] Children with paraplegia are more likely to ambulate than those with tetraplegia. [23, 24]

Two recent systematic reviews reported improvement in ambulation when a variety of LT interventions were initiated in children during the chronic phase of their injury. [5, 23] Furthermore, after the LT there were positive results for gait speed, distance and participation. [5, 23] An additional benefit resulting from upright mobility and LT is the reduced risk of developing hip dysplasia or severe scoliotic curves. [10, 23] At present, there is neither official guideline for LT or knowledge of its long term effects in the pediatric population due to lack of evidence. [5] The progression of LT depends on the segmental control and the ability of the participant to independently maintain proper trunk, pelvis, and lower extremity postural alignment. [5] Factors like increased walking speed and distance, decreased BWS, decreased manual assistance and ambulation over ground promote a more functional gait pattern. [5, 24]

Functional electrical stimulation (FES) and neuromuscular electrical stimulation (NMES) are used to promote muscle activity in adults. [3, 24] Although there is not much evidence about the use of them in children, a randomized study including 30 children with SCI aged from 5 to 13 years old reported improved muscle volume after NMES, improved muscle

volume and strength after FES cycling and no differences after passive cycling. [3]

Scoliosis

Children with SCI have high risk for developing scoliosis due to their on-going musculoskeletal growth. [3, 18, 21, 25, 26] Age at time of injury is the most important predictor for spinal curvature progression. [3, 25, 26] Patients who sustained a SCI before the adolescent growth spurt has take place have increased risk for developing scoliosis. [3, 11, 21] Indeed, children who sustained an injury under the age of 14 years are in great risk of developing scoliosis within 10 years past the injury. [25] The secondary complications of neuromuscular scoliosis often include pelvic obliquity, skin breakdown, pulmonary compromise, and functional decline. [26]

The management of trunk deformities is conventionally focused on progression prevention by bracing and realignment of the spine in the wheelchair, with close follow up at least until trunk growth plateaus. [11] Curves less than 40° have been shown to respond well to bracing [26]. Bracing in neuromuscular scoliosis with Cobb angles less than 20° delays surgical correction and in angles less than 10° may prevent scoliosis progression [3, 11]. However, thoracolumbar sacral orthosis (TLCO) treatment is reported to impair daily activities. [3, 11, 26] Tolerance in bracing is even more difficult in the presence of hyperhidrosis or in warm environments. [26]

Activating the trunk muscles and improving Segmental Assessment of Trunk Control (SATCo) scores reflective of greater trunk control may reduce the risk and incidence of scoliosis. The SATCo is a new reliable and validated pediatric measurement instrument used to assess and measure improvements in trunk control regardless of chronicity, severity and level of injury in children with SCI lacking independent sitting or impaired sitting control. [27]

Although there is very limited evidence concerning the results of surgical treatment of spinal deformity following SCI [11], children who sustain a SCI before the age of 12 years have a 3.7 times increased possibility to undergo spinal fusion. [3, 26] Surgical treatment of neuromuscular scoliosis is indicated only in patients older than 10 years and in cases where Cobb angle is greater than 40-45°, there is rapid progression of the curve, or functional problems exist [1, 3].

Hip subluxation

Hip subluxation and dislocation is the second most common orthopedic complication in children who sustain a SCI before the age of 10 years, affecting about 90% of these patients. [1] This complication may occur no matter if spasticity is present or not. [1] A recent review concluded that children under the age of 5 years with complete SCI and paraplegia have a significantly increased risk to develop hip subluxation. [10] Hip surveillance and orthopedic surgery are suggested. [1] Among 146 injured children less than 5 years old, 57% presented hip dysplasia and only 8% had surgery. [10] An abduction pillow or pommel can be useful to support hip abduction in supine and sitting position respectively. [24]

Autonomic dysreflexia

As seen in adults, pediatric patients with T6 or higher SCI are at risk of developing autonomic dysreflexia (AD). However, this life threatening complication is more difficult to be recognized in children due to their inability to express symptoms of AD. This is especially true in children younger than 5 years which rarely show any symptoms. [1] Measurement of blood pressure (BP) provides a reliable diagnosis of AD. A 15 mmHg increase of BP above baseline in children and a 15-20 mmHg increase of BP above base line in adolescents are considered to be signs of AD. [3] However, normal BP values vary with age, and are also related to body mass index (BMI) and type of SCI. Indeed, as the child grows BP increases and children with paraplegia and incomplete injury tend to have higher BP than non affected children. [3]

Removal of the inciting factor and monitoring of BP and heart rate is the initial management of AD. Bladder distention and fecal impaction are the main causes of AD. [1, 3] AD may also appear during surgical procedures. [3] Medications recommended for children are nitroglycerin and nifedipine and they are used when the systolic BP exceeds 120 mmHg in children younger than 5 years, 130 mmHg in children 6–12 years, and 140 mmHg in adolescents. [3]

Urology

Management of neurogenic bladder in pediatric SCI

patients aims in preservation of renal function, prevention of life-threatening complications and promotion of continence which correlates with the child's self-esteem and participation. [1] A recent study, including 127 children under the age of 5 years who sustained a SCI and had no bladder control, reported that 82% of children were on an intermittent catheterization program and became independent at a median age of 8 years and 4 months. [10] Clean intermittent catheterization (CIC) is the appropriate management of neurogenic bladder in children over 3 years old. The goal of CIC is for children to obtain complete independence by the age of 5 to 7 years old, given there is adequate hand function. Children with inadequate hand function should gain independence in verbal direction of care. [1] Furthermore, children with poor hand function may benefit from a continent catheterizable urinary conduit, known as Mitrofanoff procedure. [1, 28] Urodynamic studies (UDS) in children with SCI may include uroflowmetry, electromyogram, and cystometry and should be performed for the first time after the spinal shock has passed (at least 3 months after the injury). Follow-up UDS is necessary as children with SCI will need long-term evaluation as they grow and bladder function may change over time. [28]

Normal urine output is age-dependent and should adjust according to the child's growth. [3, 28] Bladder capacity in children is calculated as follows: (age + 2) × 30 = Bladder Capacity (in milliliters/cubic centimeters). [28] Bladder capacity in children with SCI has been found to be less than expected. A normal adultsize bladder does not develop approximately until the age of 10. [3, 18] Inadequate bladder capacity may result in need for impractical frequency of catheterization or incontinence. [3] Management of incontinence with oral anticholinergic medications usually is effective [3]. Studies in small patient groups have reported that daily intravesical oxybutynin and onabotulinumtoxin A injections into the bladder detrusor increased bladder capacity, but there are no specific guidelines about the administration in children with SCI. [3, 28] Studies regarding long term effects from the injections showed improvement in capacity up to 10 months and less improvement over time. [28] When conservative management fails augmentation cystoplasty can be performed. [3, 28]

Urinary tract infections (UTI) are a common problem and may require treatment when there are symptoms such as fever, chills, AD, or exacerbation of spasticity. [28] Bacteriuria is present in 70-76% of children who use clean intermittent catheterization method to empty their bladder. [28] Prophylactic antibiotics for asymptomatic bacteriuria are not recommended. [28] UTIs increase the risk for urinary stones formation and may lead to pyelonephritis and sepsis. [28] In a recent cross-sectional retrospective study, including 76 pediatric patients with traumatic and non-traumatic SCI, it was reported that approximately 43% of patients suffered from Upper Urinary Tract (UUT) deterioration. Iatrogenic trauma to the spinal cord, presence of abnormal radiological lower urinary tract findings, absence of CIC and antimuscarinic drug treatment, maximum detrusor pressure (maxPdet) greater than 70 cmH₂O during the storage phase and bladder volume ratio less than 0.7 constitute causes of UUT deterioration in children with SCI. [29]

Neurogenic Bowel

Neurogenic bowel dysfunction is common after SCI. The assessment for bowel dysfunction and neurogenic bowel management and rehabilitation in children with SCI are similar to those in adults. Neurogenic bowel might be the result of upper or lower motor neuron injury. [1, 3] Children aged from 2 to 4 years old are recommended for bowel programs. The bowel program is usually a tailored treatment plan that includes consideration of diet, physical activity level, oral or rectal medications and scheduling of bowel care. [1, 3]

In conclusion, study of the epidemiology and etiology of pediatric SCI may help in creating prevention strategies and education campaigns about safety of children and adolescents. Caring for children or adolescents with SCI presents many unique challenges such as the management of the secondary health complications. Furthermore, management of pediatric SCI should be based on normal life milestones, on changing objectives in each developmental stage, and on using appropriate strategies to facilitate adjustment and maximize independence across the spectrum of physical and emotional maturity levels.

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