

# Pediatric Spinal Cord Injury

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## **Pediatric Spinal Cord Injury**

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*To all children and adolescents  
with Spinal Cord Injury*

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## LIST OF ORIGINAL PUBLICATIONS

The thesis is based on the following publications, which are referred to in the text by the corresponding Roman numerals.

- I Augutis M, Malker H, Levi R (2003).  
Pediatric spinal cord injury in Sweden;  
how to identify a cohort of rare events.  
*Spinal Cord* 41: 337-346
- II Augutis M, Levi R (2003).  
Pediatric spinal cord injury in Sweden:  
incidence, etiology and outcome.  
*Spinal Cord* 41: 328-336.
- III Augutis M, Abel R, Levi R (2006).  
Pediatric spinal cord injury in a subset  
of European countries.  
*Spinal Cord*. 44(2):106-12.
- IV Augutis M, Levi R, Asplund K, Berg-Kelly K (2006).  
Psychosocial aspects of traumatic spinal cord injury  
with onset during adolescence – a qualitative study.  
*The Journal of Spinal Cord Injury Medicine*  
(accepted January 2007).

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## Abbreviations

SCI	Spinal Cord Injury
PedSCI	Pediatric Spinal Cord Injury
SCIWORA	Spinal Cord Injury Without Radiological Abnormalities
ASIA	American Spinal Injury Association
NSCISC	The National SCI Statistical Center Database
NIDRR	National Institute for Disability and Rehabilitation Research
PIN	Personal Identification Number
HDR	Hospital Discharge Register
CDR	Cause of Death Register
MBR	Medical Birth Register
ICD	International Classification of Diseases
ISCoS (IMSOP)	International Spinal Cord Society (International Medical Society of Paraplegia)
EU	European Union
EEC	European Economic Community
EEA	European Economic Area
WHO	World Health Organization
EURODIS	The European Organisation for Rare Diseases
PRINTO	Paediatric Rheumatology International Trials Organisation
CI	Confidence Interval
SD	Standard Deviation

## ABSTRACT

Sustaining a Spinal Cord Injury (SCI) alters every aspect of a person's life significantly and irreversibly. Since SCI of traumatic etiology predominately occurs in young adult males, most research has focused on this group. Other demographic groups, such as children and women, have received much less attention. A SCI striking a child or adolescent, i.e. "pediatric SCI" (pedSCI), presents unique challenges due to ongoing physical and psycho-social development. As long as knowledge about pedSCI remains incomplete, effective targeting of high risk groups and situations for purposes of prevention is impossible. Additionally, the organization of care and rehabilitation for pedSCI requires a solid understanding of the scope of the problem.

### *Aims*

The overall aim of this thesis is to define the incidence, management and consequences of SCI during childhood and adolescence in Sweden (**Study I, II**) and Europe (**Study III**) and, furthermore, to explore psychosocial factors of importance for rehabilitation from the patient perspective in persons who have sustained SCI during early and mid adolescence (11-15 years) (**Study IV**).

In **Study I**, a method of identifying pedSCI using population registers is described. Population registers offer a practical initial source for identification of a study population, although the number of false positives exceeds 70%. Subsequently, further refinement and quality control is necessary in order to verify a true cohort. Such further verification proved to be both time-consuming and tedious.

In **Study II**, data from population registers, County Habilitation Centers and several informal sources were used to estimate the incidence of pedSCI in Sweden during the years 1985-1996 among children aged 0-15 years. Contacts with treating hospitals, reviews of medical records and/or personal interviews were used to verify primary data. In total, 92 cases were thus identified. The incidence was found to be 4.6 cases/millionchildren/year. When excluding prehospital fatalities, the incidence was 2.4. The main cause of injury overall was traffic accidents. Among surviving adolescents, sports-related injuries were as common a cause as traffic accidents. The survivors were treated in 18 different hospitals. We conclude that pedSCI is rare in Sweden. This is likely to be a benefit of the systematic injury prevention programs that have been implemented in Sweden. On the other hand, care of these patients was found to be very fractionated and unsystematized.

In **Study III**, a short semi-structured questionnaire was sent to respondents working with SCI in 19 countries in Europe. PedSCI was reported rare throughout Europe. Only in Portugal and Sweden had the incidence of pedSCI previously been determined. The reported incidence estimates varied considerably, from about 0.9 cases/million children/year to about 23 cases/millionchildren/year. The management differed depending on differences in the division of labor between pediatric and adult health care providers and on the local organization of health care.

In **Study IV**, twenty-four of the 28 persons who had sustained a SCI in early-to-mid-adolescence in Sweden during 1985-1996 participated in the study. Semi-structured interviews were made an average of 10 years post injury. Narratives were analyzed qualitatively according to content analysis. Parents and peers were found to have played an important role in helping the persons cope with their injury. Parents frequently acted as advocates in interactions with health care providers, as supporters and as containers of sorrow, frustration and anger. Peers acted as promoters of activity and identity development. Health care providers were fairly often perceived as not sufficiently facilitating the involvement of this network. Rehabilitation professionals might be encouraged to increase their knowledge of adolescence medicine in order to better meet the specific needs and demands of persons in this age group. It is further suggested that parents and peers be considered important partners in the joint rehabilitation effort.

**Keywords:** *Pediatric; Spinal Cord Injury; Epidemiology; Adolescence; Psychosocial; Rehabilitation.*



## SAMMANFATTNING

En ryggmärgsskada förändrar drastiskt och ofta varaktigt olika aspekter av en persons liv. Eftersom en ryggmärgsskada är vanligast bland unga män (16-30 år), har också forskningen varit inriktad mot denna grupp, medan andra sub-grupper som barn och kvinnor har fått mindre uppmärksamhet. Hos barn och ungdomar manifesterar sig skadan annorlunda än hos vuxna då de befinner sig under en ständig utveckling både fysiskt och psykosocialt. Kunskap om förekomst av, och orsaker till, ryggmärgsskada i denna grupp är nödvändig för att identifiera riskgrupper, förbättra förebyggande åtgärder samt för att planera vård och rehabilitering

### **Syfte**

Det övergripande syftet med avhandlingen är att utröna förekomst, vård och konsekvenser av traumatisk ryggmärgsskada bland barn och ungdomar i Sverige (**I, II**) och Europa (**III**). Vidare, att undersöka de psykosociala faktorer som har upplevts befrämja eller hindra rehabiliteringsprocessen bland personer som förvärvade ryggmärgsskadan under ungdomsåren (tidig och mellan-adolescens) (**IV**).

I **Studie I** beskrivs en metod för att identifiera ryggmärgsskada bland barn och ungdomar genom att använda befolkningsregister. Dessa register är användbara för en första identifikation av studiepopulationen. Dock visade fortsatt kvalitetssäkring att över 70% av fall med ryggmärgsskadediagnos i registren var falskt positiva. Följaktligen måste data ur dessa register ytterligare granskas för att säkra kvalitet och validitet. Denna vidare verifiering var tidskrävande och omständlig.

I **Studie II** används data från befolkningsregister, länshabiliteringar och informella källor för att uppskatta fall av ryggmärgsskada bland barn och ungdomar (0-15 år) i Sverige mellan åren 1985-1996. Kontakt med behandlande sjukhus, journalgenomgång och/eller personlig kontakt användes för att verifiera data. Totalt 92 fall identifierades. Incidensen var 4.6 fall/miljoner barn/år. Efter exkludering av fall som avlidit innan ankomst till sjukhus var incidensen 2.4. Den vanligaste orsaken till skada var trafikolyckor. Bland överlevande ungdomar var sportrelaterade skador lika vanliga orsaker som trafikolyckor. De som överlevde skadan vårdades vid 18 olika sjukhus. Ryggmärgsskada bland barn och ungdomar under 16 år är således ovanlig i Sverige. Studien visar också att vård och rehabilitering för denna grupp är spridd och osystematiskt organiserad.

I **Studie III** skickades en enkät till respondenter som arbetar med ryggmärgsskaderehabilitering i 19 länder i Europa. Endast i Portugal

och Sverige är incidensen av traumatisk ryggmärgsskada bland barn och ungdomar fastställd. Den rapporterade incidensen varierade mycket, från ca 0.9 fall/miljoner barn/år till ca 23 fall/miljoner barn/år. Generellt är ryggmärgsskada i denna grupp ovanlig i Europa. Vård och rehabilitering skiljde sig mellan länderna bland annat som en konsekvens av skillnader i arbetsfördelning mellan pediatrikisk vård och vuxenvård, samt även som en konsekvens av övergripande skillnader i hälso- och sjukvårdsorganisation.

I **Studie IV** deltog 24 av de 28 personer som ryggmärgsskadades i åldern 10-15 år i Sverige under åren 1985-1996. Semistrukturerade intervjuer genomfördes under 2002–2004, i genomsnitt 10 år efter skadetillfället. Intervjumaterialet analyserades kvalitativt enligt så kallad innehållsanalys. Studien visade att föräldrar och kamrater spelar en betydande roll för personernas återanpassning. Föräldrarna hade ofta roller som talesmän, supportrar samt hållbarare av sorg, frustration och ilska. Kamraters närvaro och engagemang befrämjade aktivitets- och identitetsutveckling. Mot detta stod beskrivningar av att vårdpersonalen i många fall inte hade understött delaktighet av ungdomarnas sociala nätverk under rehabiliteringen. Vård och rehabilitering av ryggmärgsskadade barn och ungdomar bör i högre grad underlätta engagemang av den unges sociala nätverk, speciellt föräldrar och kamrater, liksom ta hänsyn till åldersspecifika utvecklingsaspekter.

## Personal introduction

*This thesis originated from some clinical experiences that deeply touched me. Questions pressed to be answered, to be put back into real life, for the benefit of children and adolescents with SCI.*

Many persons have asked me why I embarked on this project. This is understandable, since during the more than thirty years I have dedicated to being a physiotherapist, I have only had four children with pedSCI as my patients!

My everyday workplace is the department of Pediatrics of a county Hospital, without any specialized rehabilitation for children with disabilities.

However, since 1982, I have been an active member of a non-profit organization, *Rekryteringsgruppen*, which arranges, among other things, training camps promoting sports as a tool for rehabilitation and empowerment for persons with disabilities. *Rekryteringsgruppen* uses a role model program, involving persons with disabilities (mainly SCI) to act as leaders in the organization and on the camps. *Rekryteringsgruppen* has links with most SCI units in Sweden, and they continually try to recruit recently injured persons to the organization. Through *Rekryteringsgruppen* I have met many recently injured persons with SCI from all parts of our country. I have also worked in the international branch of this organization, aiming at implementing their program in Eastern Europe, e.g. in Poland, Ukraine, Lithuania and Belarus.

In 1988 I had the opportunity to spend four months at the Austin Hospital in Melbourne to learn more about acute care and early rehabilitation of persons with SCI.

In the mid-1990s two children sustained a SCI in my hometown Sundsvall, and were admitted to our hospital. At that time, there was virtually no previous experience locally or regionally to rely on. I felt the frustration of not being able to provide what had to be provided, medically and psychologically, to these children and their families.

Through my work in *Rekryteringsgruppen* I have realized the great importance for patients meeting other persons with similar disabilities. Unfortunately, this was not possible to arrange at that time. Out of this frustration grew a curiosity and a determination to ponder these issues further. One friend of mine, Kerstin Wahman, encouraged me to embark on this work...I contacted Claes Hultling, who in turn persuaded Richard Levi into becoming my supervisor...and this is the result...

## INTRODUCTION

Sustaining a Spinal Cord Injury (SCI) alters every aspect of a person's life significantly and irreversibly. Since SCI of traumatic etiology predominately occurs in young adult males, most research has focused on this group. Other demographic groups, such as children and women, have received much less attention. A SCI striking a child or adolescent, i.e. "pediatric SCI" (pedSCI), presents unique challenges due to ongoing physical and psycho-social development. As long as knowledge about pedSCI remains incomplete, effective targeting of high risk groups and situations for purposes of prevention is impossible. Additionally, the organization of care and rehabilitation for pedSCI requires a solid understanding of the scope of the problem.

## BACKGROUND

### Epidemiology

Epidemiology is the study of the distribution, causes and determinators of disease frequency. Two fundamental assumptions underlie the study of epidemiology. Firstly, human disease does not occur at random; and secondly, human disease is linked to causal and preventive factors which may be identified through systematic investigation of different populations or subgroups of individuals in different places and different times <sup>1</sup>.

Vast amounts of data are available for clinical and epidemiological research through various administrative information systems.

Analyses of population-based administrative registers are practical and cost-effective strategies to utilize already existing data for epidemiological purposes<sup>2</sup>. However, there are caveats when using such databases for research purposes <sup>3</sup>. Since the primary purpose of many registers is administrative rather than scientific, this may reflect negatively on data quality <sup>4</sup>.

The country of Sweden has an unusually long tradition of population registration, dating back to 1749 when "*Tabellverket*" <sup>5</sup> was founded. Since then, a large number of nationwide population-based disease or effect registers have been developed <sup>4</sup>. The drop-out rates are typically very low, usually less than 5%. The registers are frequently used for research, evaluation, planning and other purposes by a variety of users <sup>6</sup>.

Furthermore, there exists a unique system of personal identification number (PIN) for every Swedish resident <sup>6</sup>.

The registers and PINs, together with modern computer technology, facilitate linkage of exposure and outcome data at the individual level <sup>4</sup>.

In the literature there are many cautions about surveys based on register studies, pointing to biases in the reporting methods, data collection systems and classification patterns <sup>7</sup>. The use of discharge diagnoses for identification of cases, for example, may be problematic due to variations in coding procedures, coding errors, incomplete coding, lack of specificity in available codes and clinical errors in diagnosis <sup>2</sup>.

Data collection of rare events (such as SCI) from population-based registers poses particular problems due to the large amount of data that has to be screened in order to obtain a study population large enough to enable conclusions. The implication is that verification of every single case must be made very carefully to avoid misclassifications and/or differences in classification.

In many countries population based registers do not exist and estimates of diseases or injuries are based on either retrospective data from hospitals <sup>8</sup> or prospective estimates attempting to capture all cases within a geographic area <sup>9,10</sup>.

The validity of incidence estimates is obviously dependent on how well true cases are recognized, identified and verified.

Knowledge of the incidence and etiology of SCI is of importance in order to identify risk groups, improve prevention and for planning of care and rehabilitation. Although SCI indeed is a rare event, it is nevertheless important from a societal perspective due to its medical and socio-economic consequences. Thus, there is a need for improved registration of SCI with uniformity in methodology <sup>11</sup>.

The development of the Model Spinal Cord Injury Programs, in addition to systematic trauma registration in the National SCI Statistical Center database (NSCISC) in the USA has greatly contributed to our current knowledge of SCI <sup>12</sup>.

In order to standardize and to improve the collection and reporting of SCI a committee of experts has recently developed an International Spinal Cord Injury Core Data Set <sup>13,14</sup>.

## Incidence

The annual incidence of SCI throughout “developed” countries is between 15-40 cases per million <sup>15</sup>. SCI can occur at any age, the effects are usually permanent and currently there is no cure <sup>16</sup>. NSCISC data indicate that more SCIs occur in the 16-30 age group than in all other age groups combined. The average age at injury is 30.7 years and the most frequent age at injury is 19 years. Survival rates have increased and many patients live into old age. Consequently, the lifetime cost of care will be high<sup>16</sup>.

The incidence of traumatic SCI in Sweden is about 10-15 persons/million inhabitants per year <sup>17</sup>, i.e. around 120 persons sustain a SCI each year.

Data specifically regarding pedSCI is scarce (see Table 1).

During the last decade, especially in the USA, pedSCI have received increasing scientific interest. However, children with this injury, in particular the youngest, typically are treated in children’s hospitals rather than in adult spinal units (personal communication, 24 January 2001) <sup>18</sup> and are therefore not captured in the NSCISC database.

A second database was developed in 1987, to address the lack of pedSCI data. This database comprises data from 1973 onward from three SCI units of the Shriners Hospitals for Children (Philadelphia, Chicago, and Sacramento), and uses a similar data set as NSCISC <sup>19</sup>.

.

**TABLE 1.** The estimated incidence of pedSCI reported from the USA, Canada, New Zealand, the Netherlands and Portugal.

<i>References</i>	<i>Place</i>	<i>Study years</i>	<i>Age-groups (years)</i>	<i>PedSCI/million children/year</i>	<i>Death prior to hospital admission included (Yes/No)</i>	<i>Estimated overall incidence of traumatic SCI/million population/year</i>
Surkin <i>et al</i> 2000 <sup>10</sup> .	Mississippi, USA	1992-1994	0-14	11.3	Y	77
Buechner <i>et al</i> 2000 <sup>20</sup>	Rhode Island USA	1994-1998	0-14	9	N	56
Martins <i>et al</i> 1998 <sup>21</sup>	Portugal (central region)	1998	0-14	27	Y	57.8
Warren <i>et al</i> 1995 <sup>22</sup>	Alaska USA	1991-1993	0-14	14	Y	83
Thurman <i>et al</i> 1994 <sup>23</sup> (*personal communication -2001-01-16) <sup>24</sup>	Utah USA	1989-1991	0-14	*16.7 <sup>24</sup>	Y	43
Price <i>et al</i> 1994 <sup>25</sup>	Oklahoma, USA	1988-1990	0-14	6	N	40
Woodruff and Baron 1994 <sup>26</sup>	West Virginia USA	1985-1988	0-14	1.0	N	
Acton <i>et al</i> 1993 <sup>27</sup> (*Personal communication 2002-09-11) <sup>28</sup>	Arkansas, USA	1980-1989	0-14	*4.98 <sup>28</sup>	N	33.18 (1989) Carol <sup>29</sup>
Kewalramani <i>et al</i> 1980 <sup>30</sup>	California USA	8 years	1-15	18.2	Y	
				8.9	N	
Dixon <i>et al</i> 1993 <sup>31</sup>	New Zealand	1979-1988	0-14	8.6	N	43.3
		1988	0-14	16	N	49.1
Van Asbeck <i>et al</i> 2000 <sup>32</sup>	Netherlands	1994	0-10	N=2	N	10.4
Pickett <i>et al</i> 2006 <sup>33</sup>	Canada	1997-2001	0-14	3.37	N	41.79
Vitale <i>et al</i> 2006 <sup>34</sup>	USA	1997-2000	0-18	19.9	N	

Due to differences in case definitions, case ascertainment and age limits as to what is considered “pediatric” as opposed to “adult” SCI, comparisons of incidence and external causes are problematic <sup>27,30</sup>. In particular, the inclusion of patients aged 15 and above in case definitions of pedSCI will substantially inflate the incidence rate for the pediatric group due to the incidence peak at 15-24 years <sup>23,26</sup>.

The pedSCI population is very heterogeneous, both as regards epidemiological characteristics within different age groups and as compared to adult-onset SCI. For example, the massive male dominance as is seen in adults, is not present in the youngest age <sup>19,35</sup>.

Therefore the epidemiologic study of pedSCI has to include developmentally based age-at-injury groupings in order to capture the differing psychological and behavioral characteristics from infancy through adolescence <sup>19</sup>.

### **Cause of injury**

In the USA most cases of pedSCI, particularly in the youngest age group, occur in motor vehicle accidents <sup>19,30,36-39</sup>.

The correct use of safety devices such as seat belt restraints and booster seats has decreased morbidity and mortality in traffic accidents among children. However, lap-belt induced injuries may occur when a small child (4 to 9 years of age) outgrow the child safety seats designed for younger children and are restrained in seat belts designed for adults. The lap belt raises above the pelvic brim, resulting in flexion and distraction forces acting on the mid-lumbar spine, the abdominal wall, and the viscera <sup>40,41</sup>. Up to the age of approximately 4 years, it is safest for a child to travel facing rearwards. A small child needs an infant seat, child safety seat or booster seat/cushion. Not until the age of 10-12 years can a child travel safely using a regular seatbelt <sup>42</sup>.

Sports-related injuries <sup>19,38,43,44</sup> and violence are other common causes of injury during adolescence according to US data <sup>19,45</sup>. Causes unique to pedSCI include birth injury (e.g. forceful traction and angulation of the spine during delivery) <sup>46-49</sup>, child abuse (e.g. aggressive shaking leading to C2-C3 subluxation) and C1-C2 subluxation associated with tonsillitis or pharyngitis. Children with preexisting disabilities such as achondroplasia, juvenile rheumatoid arthritis and Down syndrome run a higher risk for SCI <sup>35</sup>.



## Medical aspects

The pathophysiology of traumatic SCI is complex. The tissue damage is due to primary and secondary events. Primary injury refers to the mechanical disruption of axons as a result of stretch or laceration, for which no current treatment exists<sup>50</sup>. Secondary injury is due to a large number of factors that lead to progressive spinal tissue damage during a period of time after the primary event<sup>50</sup>. Intensive research efforts are directed towards limiting secondary injury<sup>17</sup>.

SCI results in impairment or loss of function due to damage of neural elements within the spinal canal in the cervical, thoracic, lumbar or sacral segments of the spinal cord. By convention, damage to the cauda equina is also included in the concept of SCI. Clinically, SCI is reflected in<sup>17,51,52</sup>:

- Impairment or absence of voluntary motor activity below the level of lesion (i.e. paresis or paralysis).
- Impairment or absence of sensory function below the level of lesion.
- Impairment or absence of volitional control of bladder and bowel emptying, thus leading to incontinence and/or retention.
- In suprasacral spinal cord lesions reflexogenic movements and muscular hypertonicity as manifestations of spasticity (i.e. the upper motor neuron syndrome).
- In lesions of conus medullaris and cauda equina the presence of flaccid paresis (i.e. the lower motor neuron syndrome).

Additionally, SCI leads to varying degrees of impairment of autonomic functions as well as metabolic and hormonal derangements of potential pathogenic significance<sup>17,50</sup>.

For purposes of standardizing the assessment of the level and completeness of an SCI, a protocol was developed by the American Spinal Injury Association (ASIA) in 1982 (ASIA Standards for Classification of Spinal Injured Patients)<sup>52</sup>. The latest revision was made in 2003<sup>51</sup>. The utility of these standards for pedSCI has been shown to be poor, in particular as regards the precision of the motor exam in children under 15 years of age and the sensory exams in children under 5 years of age<sup>53</sup>.

The following terminology is utilized universally<sup>52</sup>:

- **Tetraplegia** - Injury to the spinal cord in the cervical region with associated loss of muscle strength in all 4 extremities.
- **Paraplegia** - Injury in the spinal cord in the thoracic, lumbar, or sacral segments, including the cauda equina and conus medullaris. With associated loss of muscle strength in the lower extremities
- **Complete injury** - complete absence of sensory or voluntary motor function in the lowest sacral segment (S4-5).
- **Incomplete injury** – partial preservation of sensory and/or motor functions below the neurological level including the lowest sacral segments.

SCI leads to impairment of function in many organ systems, and also to a persisting vulnerability for several medical complications. The management thus requires attention to multiple body systems<sup>54</sup> (Table 2):

**TABLE 2.** Overview of traditional management of spinal-cord injury. Reprinted from *The Lancet*, Vol. 359, McDonald J, Sadowsky C, *Spinal-cord injury*, Pages 417-25, Copyright (2002), with permission from Elsevier<sup>54</sup>.

Medical	Surgical	Rehabilitative
Spinal stabilisation: spine immobilisation during transport and resuscitation <b>Cardiovascular:</b> haemodynamic instability; autonomic dysfunction; thromboembolism <b>Respiratory system:</b> respiratory failure; atelectasis; pneumonia; vent-dependent care <b>Gastrointestinal system:</b> ileus; impaction, constipation; gastric and duodenal ulcers; GORD; cholelithiasis <b>Genitourinary system:</b> urinary tract infection; hydronephrosis; cycto/nephrolithiasis  <b>Dermatological:</b> pressure ulcers  <b>Musculoskeletal system:</b> osteoporosis; heterotopic ossification; fractures; overuse syndromes; acute and chronic pain	Internal fusion/instrumentation; external orthoses  Tracheostomy  Urinary system augmentation; diversion procedures; penile implants; lithotripsy; sphincterotomy Pressure ulcer repair  Treatment of delayed neurological and spine complications: syringomyelia; focal nerve entrapments; central pain, spasticity; spinal instability; implantation of intrathecal drug-delivery systems.	Management of chronic haemodynamic issues: autonomic dysreflexia  Preventive respiratory care; respiratory conditioning programme  Establish predictable bowel continence programme; preventive gastrointestinal care Programme to establish bladder continence; preventive genitourinary care; sexual dysfunction programme Establish skin integrity programme; prevent and manage pressure ulcers Prevent/manage musculoskeletal complications: contractures; spasticity; postural abnormalities; skeletal deformities; long-term intrathecal drug treatment  Functional retraining in self-care; mobility; psychosocial adaptation; vocational and recreational skills; adaptive equipment and orthotic devices
GORD=gastro-oesophageal reflux disease.		

Certain biological aspects distinguish pedSCI from adult-onset SCI. The anatomy and biomechanics of the immature cervical spine differs from that of the adult. The injuries sustained by children also vary with age. Young children (less than 8-9 years of age) are vulnerable to flexion forces to the upper cervical spine; there are fewer fractures and subluxations than in children above 9 years of age. The syndrome of Spinal Cord Injury Without Radiographic Abnormality (SCIWORA) is more frequent in the young age group and the neurological lesion is often severe. Delayed onset of neurological damage is reported to sometimes occur with a latency period of 30 minutes to 4 days<sup>55</sup>.

In a registry study over the past 3 decades comprising 35,000 persons with pedSCI, a higher percentage of paraplegia in the youngest age groups 0-5 years and 6-12 years was found. Upper cervical injuries were least common in the youngest age group<sup>19</sup>.

Adolescents tend to sustain injuries similar in pattern to those of adults<sup>43,56</sup>.

The acute somatic care of pedSCI is in many respects similar to that of adult-onset SCI. Bowel and bladder regimes, skin care to prevent decubital ulcers, physical therapy to prevent atrophy and contractures, attention to autonomic dysreflexia, and careful pulmonary care exemplify measures to be considered<sup>57</sup>.

Hypercalcemia is seldom symptomatic in adult SCI patients, but has to be considered in pedSCI<sup>57,58</sup>. There is an increased incidence of this condition in older children and adolescents, particularly males, due to the combination of a proportionally large bone mass, rapid growth-related bone turnover and increased bone resorption associated with immobilization<sup>57</sup>. Another particular risk associated with pedSCI is the development of latex allergy<sup>57</sup>.

Infants and younger children with tetraplegia are at particular risk of incipient respiratory failure, which may present as sleep apnea<sup>57,59</sup>.

Late spinal deformity after spinal injury may occur in children and often appears at times of growth spurts. Scoliosis is reported to have developed in 97% of 130 patients who were injured before the adolescent growth spurt<sup>60</sup>. Young age is also associated with the development of hip luxation<sup>61</sup>.

Better survival after a SCI correlates with a lower neurological lesion level and incompleteness of the injury. Pneumonia, pulmonary embolism, and septicemia are the most common causes of death after acute injury in SCI in all ages<sup>62</sup>. Suicide and substance abuse have been reported as

being additional and potentially preventable causes of death among adolescents and young adults after SCI <sup>63-65</sup>.

SCI often occurs together with extraspinal injuries. Most common are injuries to the head, extremity fractures and/or major chest and abdominal injuries <sup>56,66</sup>.

A recent review has reported that over a period of 30 years there has been a 40% decline in mortality during the first 2 years after injury. However, despite developments in long-term rehabilitative care, improvements in the life span of persons with SCI beyond the post 2 - year period was not found <sup>67</sup>.

## **Psychosocial aspects**

### ***Children***

Children are not “small adults” but have unique medical needs and require different psychological and social support than adults <sup>68</sup>. Their ability to understand and cope is influenced by age, developmental stage and experiences <sup>69,70</sup>.

Care of very young children is complicated by their restricted abilities to localize and verbalize symptoms. They often need help from their parents to express their needs. The clinical manifestations of e.g. autonomic dysreflexia and respiratory failure may be subtle <sup>68</sup>.

Developmentally appropriate information to children is of outmost importance, helping to clarify misconceptions and fears <sup>71</sup>.

Properly age-adapted care can positively influence the degree of cooperation, as well as the long-term experiences of care. Play and age-appropriate communication may be used to present information and to prepare the child for medical events or procedures <sup>72</sup>.

### ***Adolescents***

Adolescence, i.e. the period between 10-21 years of age <sup>73</sup>, is a transitional period from being dependent within a family to becoming an independent person. It includes the gradual adaptation to a variety of emerging social and emotional responsibilities. Adolescents have to adapt to the gradual physical development into an adult body and consequently incorporate a “new” body image. Most important, the young person will typically experiment with a variety of life styles and move in and out of different circles of peers and important adults in order to ultimately secure a stable identity <sup>74</sup>. Consequently, adolescence is the key period for learning “adulthood” <sup>75-79</sup>.

Sexuality is a core issue in adolescence. There is a marked increase in curiosity and interest during this period <sup>80</sup>. Romantic relationships typically appear and provide many aspects of role identification and lifestyles <sup>77</sup>.

Parents continue to be important throughout adolescence. They ideally provide emotional security, guidance and discussions, and serve as a safe recipient for the acting out of teenage rebellion. Adolescent development places a great deal of stress on parents to be flexible. They have to adjust their parenting styles and adapt to the increasing independence of their children <sup>81,82</sup>.

The World Health Organization (WHO) has described the clinically most relevant features of adolescence as a part of a so-called orientation programme <sup>73</sup>. The program was created with the goal to improve and adapt health care services to young people's special needs and concerns. Its key features are depicted in Table 3.

**TABLE. 3.** Characteristics of phases of adolescence. *Orientation Programme on adolescent Health for Health-Care Providers, WHO* <sup>73</sup> (Published with permission).

Category of change	EARLY 10-13 to 14-15 years	MIDDLE 14-15 to 17 years	LATE 17-21 years (variable)
Growth	Secondary sexual characteristics appear. Growth accelerates and reaches a peak.	Secondary sexual characteristics advanced. Growth slows down, approximately 95 % of adult stature attained.	Physically mature.
Cognition	Concrete thinking. Existential orientation. Long-range implications of actions not perceived.	Thinking is more abstract. Capable of long-range thinking. Reverts to concrete thinking when stressed.	Established abstract thinking. Future-oriented. Perceives long-range options.
Psychosocial	Preoccupied with: rapid physical growth, body image, disrupted change.	Re-establishes body image. Preoccupation with fantasy and idealism. Sense of all-powerfulness	Intellectual and functional identity established.
Family	Defining boundaries of independence/dependence.	Conflicts over control.	Transposition of child-parent relationship to adult-adult relationships.
Peer group	Seeks affiliation to counter instability.	Needs identification to affirm self image. Peer group define behavioural code.	Peer group recedes in favour of individual friendship.
Sexuality	Self exploration and evaluation.	Preoccupation with romantic fantasy. Testing ability to attract opposite sex.	Forms stable relationships. Mutuality and reciprocity. Plans for future.

When major injury or illness occurs during adolescence, it will lead to unique challenges due to the concurrent physical and psychosocial development processes<sup>57,83,84</sup>.

Principles of psychosocial rehabilitation during adolescence are, to some extent, generic, i.e. irrespective of diagnosis<sup>85</sup>. The substantial literature on chronic illness during adolescence underscores the importance of giving due attention to the biological, emotional and social needs of the young person<sup>84,86-91</sup>. Furthermore, the social context is yet a key factor that is easily underestimated<sup>92-97</sup>.

SCI is a devastating and life-altering event<sup>57</sup>. The importance of age-related issues, and the need for developmentally based care for SCI occurring in children and adolescents, has been elaborated in several articles and studies<sup>48,57,68,98-105</sup>. Research, however, has mostly been focused on adjustment later in life<sup>106-109</sup>, and on the transition to adult care settings<sup>110-112</sup>, rather than on the primary rehabilitation process and the early readjustment thereafter.

Only few studies have specifically focused on the psychosocial aspects of care, from the perspective of young persons themselves<sup>89,90,94-96,113</sup>.

## Rehabilitation

Until World War II, over 80% of patients with SCI died within a few weeks after the injury, mainly due to infected pressure ulcers and urinary tract infections and renal failure<sup>114</sup>.

A new era of comprehensive rehabilitation began when, in the 1940s, Donald Munro in the US, and Ludvig Guttman in the UK started SCI units. This pioneering work has been followed by other countries.

Life-long follow-up is necessary to prevent and detect complications<sup>48,115-117</sup>. These circumstances stress the importance of improving rehabilitation programs to assist patients in reclaiming control over their situation.

SCI means lifelong disability with great costs for society due to the high consumption of care<sup>118</sup>. It is of great importance for both the individual and society that there is good quality of care and rehabilitation<sup>37,119</sup>. A multi-disciplinary team with specific training and experience is needed, in order to prevent complications and shorten hospitalization times thereby giving the patient the possibility of achieving maximum independence, optimal quality of life and good health<sup>48,99,120</sup>. It has been shown that an individual with SCI will benefit of being treated in a specialized unit as compared with a non-specialized hospital setting<sup>121</sup>.

### ***The Model SCI Systems***

The National Institute for Disability and Rehabilitation Research; NIDRR proposed a new service delivery model for SCI launched in the late 1960s. This model included: 1. Rapid case finding and referral. 2. Early rehabilitation coordinated by a specialized team. 3. A mechanism for using all essential community resources to facilitate successful rehabilitation. 4. A long-term follow-up program to ensure that gains achieved during rehabilitation were maintained<sup>16</sup>.

There are five clinical service components of a Model System:

1. Emergency medical services
2. The traumatology-intensive/acute care phase.
3. Comprehensive medical rehabilitation.
4. Psychosocial and vocational rehabilitation and services.
5. Long-term health maintenance and medical follow-up.

Other essential features of the Model SCI Systems include accessibility to care, co-ordination of services, adequate volume of patients, research and evaluation, education and training, and community outreach and advocacy<sup>16</sup>.

### ***Shriners Hospitals***

Shriners Hospitals for Children is a network of 22 pediatric hospitals in the U.S., Canada and Mexico. In 1980, Shriners Hospitals opened the first of three SCI Rehabilitation Units for children. These units, at the Chicago, Sacramento and Philadelphia locations, are among the few in the United States (and globally) specifically designed to treat children and adolescents with SCI. They also conduct research, and arrange courses and conferences regarding pedSCI<sup>122</sup>.

Three basic rules for the management of pedSCI have been proposed<sup>98</sup>:

1. To treat the whole family; 2. To use a multidisciplinary approach; and
3. To view treatment as an ongoing dynamic process that must change as the child grows and develops.

### *The SCI care in Sweden*

The provision of acute SCI care and rehabilitation in Sweden varies considerably between regions <sup>123</sup>.

Six university hospitals host specialist departments e.g. neuro-surgical and neuro-rehabilitation units.

There are still persons with SCI in Sweden who are treated in smaller county hospitals or in local general rehabilitation settings. This issue has been discussed extensively. In a work group report in 1992, initiated by the National Board of Health and Welfare, a forceful proposal was made for further centralization of SCI care and rehabilitation <sup>123</sup>. The question is, alas, still not resolved and the discussion continues <sup>124</sup>.

During the time under survey in **Study IV**, adolescents with SCI were typically treated in adult neuro-surgical and neuro-rehabilitation units during initial care and rehabilitation. This hospital-based treatment was supplemented with so-called role-model programs and training camps provided by patient associations <sup>125,126</sup>. Many camp leaders were themselves disabled, thus acting as role models showing how to manage daily living activities <sup>125</sup>.

### *Habilitation*

Children with SCI in Sweden may, after initial care and rehabilitation, be referred to a nationwide organization of Habilitation Centers. Typically, children and adolescents with congenital or neonatally acquired conditions are enrolled by the Habilitation Centers <sup>127</sup>. The Habilitation Centers are open to children up to 18 years of age with a wide variety of injuries and disabilities. During 1980s, in Sweden as well as in many other western countries, there was a shift from regional or national pediatric habilitation/rehabilitation institutions to small, multi-disciplinary teams working on a local basis, close to the child's home and school, predominantly on an outpatient basis. This shift has generally been considered beneficial. More emphasis could thereby be put on contextual factors, thus promoting empowerment and societal participation. On the other hand, decentralization has inevitably reduced the possibility to provide specialized services. Specialized rehabilitation for children and adolescents can consequently only be offered in a few major cities. Such centers are focused mainly on traumatic head injuries <sup>128,129</sup>.



## Outcome

“The goal of initial rehabilitation is to return patients to their community as quickly as possible and to assure that they are prepared to participate again in most of their typical activities. The long-term goal is to assure that patients keep pace with peers and that they are prepared to transition to a satisfying and productive adult life”<sup>98</sup>.

There are studies showing that persons with pedSCI score a good quality of life as compared to persons with adult-onset SCI<sup>109,130,131</sup>. However there are also reports of less community participation, lower life satisfaction, and lower perceived physical health as compared to healthy peers<sup>111</sup>.

High life satisfaction in adults with pedSCI has been shown to be associated with higher functional independence, better perceived mental health, higher degree of participation in the community, fewer medical complications (pressure ulcers, urine tract infections, and pain), higher marriage frequency, more often living independently (i.e. living away from parents) and being employed<sup>112</sup>.

## Prevention

Injuries caused by road traffic are major public health problems all over the world. Even if the number of deaths in Sweden resulting from road traffic is low compared with other countries; it is nevertheless the primary cause of death among children and young people. Traffic accidents also result in serious morbidity and permanent disability among young people<sup>132</sup>.

Safety promotion efforts in Sweden have focused on building safety into the environments in which children and young people live and act. This effort as a matter of course cannot prevent all accidents and emphasis has also been put on influencing behavior by laws and regulations and/or information and education. The Child Safety Commission in Sweden emphasizes that consultation with children and young people should become a natural part of the work of many agencies and programs<sup>132</sup>. Most Swedish injury prevention efforts so far have focused on young children, making adolescents a “forgotten group”. There is currently no national coordination or methodological development of preventative efforts regarding traumatic injuries<sup>132</sup>.

Prevention efforts tailored for particular stages of growth and development are needed<sup>133</sup>. Furthermore, SCI prevention in children and adolescents would benefit from guidance by epidemiologic data with respect to age, risk of sustaining an SCI, and etiologies<sup>19</sup>.

## AIMS

### General aim

The general aim was to appraise the incidence, management and consequences of SCI during childhood and adolescence (pedSCI) in Sweden (**Study I, II**) and Europe (**Study III**). Further, in **Study IV** to survey psychosocial factors of initial care and rehabilitation from the patient perspective in persons who had sustained a SCI during early and mid adolescence (11-15 years).

The studies have pursued the following specific objectives:

### *Specific aims*

#### *Articles Specific aims*

- I To define a method of identifying and verifying a population-based cohort of rare events, in this case pedSCI.
- II To appraise incidence, etiology and early outcome of pedSCI in Sweden during 1985-1996 in order to identify high-risk groups and high-risk situations.
- III To map the incidence and systems of care for pedSCI, including prevention, initial care and follow-up in a subset of European countries.
- IV To explore psycho-social factors facilitating and impeding the rehabilitation process in persons who sustained a SCI during early and mid adolescence (11-15 years).

## MATERIALS AND METHODS

### Study designs

**Study I** is a register study corroborated by medical records, personal contacts and, in some cases, clinical examination. Cases were identified in seven steps, comprising (1) definition of selection criteria, (2) the combined use of several data sources and (3) the use of various methods for verifications. The study is a “meta-study” in the respect that its scope is the description of a *method* for subject identification who have sustained rare medical events.

**Study II** is a retrospective descriptive study that provides the factual outcomes of a survey performed according to the method described by Study I, i.e. the number and characteristics of children aged 0-15 years in Sweden, who during the years 1985-1996 sustained a SCI.

**Study III** is a postal survey. A short semi-structured questionnaire was sent to respondents working with SCI in 19 European countries.

**Study IV** is a qualitative interview study. Twenty-four of the 28 persons who sustained a SCI in Sweden during 1985-1996 as adolescents (11-15 years) participated in the study. Semi-structured interviews were made during 2002 – 2004, an average of 10 years post injury. Narratives were qualitatively analyzed according to content analysis<sup>134</sup>.

### Data collection

#### *Registers*

For **Study I and II** a number of national registers (the Hospital Discharge register (HDR), the Cause of Death register (CDR), the Medical Birth register (MBR), local registers (Habilitation) and informal sources were used. Table 4 gives an overview of these sources, quality control procedures and the yield from each source.

In total, about 3.6 million possible events during 1-1-1985 until 31-12-1996 were included in the screening (Table 4).

**TABLE 4.** Sources of information, description, and quality control within the data source, total registered cases (0-15 years), and number of cases registered within each source with SCI.

<i>Sources of information</i>	<i>Description</i>	<i>Quality control within the data source</i>	<i>Total registered cases (children 0-15 years)</i>	<i>Number of cases registered with SCI</i>
<b><u>Nation-wide population registers</u></b>				
<b>The Hospital Discharge Register (HDR)</b> <sup>6, 135</sup>	Data on individual patients who have been treated as in-patients. Compulsory in Sweden since 1984	A check is made on all registrations in the HDR that all data are reported and valid e.g. personal identification number, hospital and main diagnosis. The dropout rates are very low, usually less than 4,5%. Diagnoses might be wrong for about 10 percent of the cases with a large variability between different diagnoses	Includes data for around 136.000 cases/year under the age of 16 years.  (In total 1.6 million cases were screened for SCI over a 12-year period.)	<b>249 cases</b>
<b>The Cause of Death Register (CDR)</b> <sup>6, 136, 137</sup>	All deaths in Sweden are reported to the CDR. Upon each death, a qualified physician must issue a death certificate before the person can be buried, this means that additional information i.e. a pathology report can be received after the death certificate is issued	Faults by coding are less than 3%, and the underreporting to the CDR is negligible <sup>6</sup> .	The mortality among Swedish children aged 0-15 years is around 600 cases/year.  (Over a 12-year period, around 7.200 cases were screened for SCI.)	<b>57 cases</b>
<b>The Medical Birth and Malformation register (MBR)</b> <sup>138</sup>	Includes injuries due to birth. The register is based on copies of the medical records used at antenatal care, delivery care and at the examination of the newborn infant.	The register information is checked each year with the corresponding information on infants in the civil registration. The under-reporting is about one per cent each year <sup>6</sup> . The accuracy of the diagnosis is not due to any routine quality control or follow-up.	Around 100.000 children are born yearly in Sweden and included in the register.  (Around 1.2 million cases were screened for SCI during 12 years.)	<b>42 cases</b>
<b><u>Local or regional registers</u></b>				
<b>Habilitation</b> <sup>127</sup>	Children between the age 0-18 years with chronic SCI in Sweden may, after the initial care and rehabilitation be referred to a nation-wide organization of Habilitation Centers. Usually, the Habilitation center consists of many small multi-disciplinary teams who work on a local basis, close to the child's home and school.	The registers are new and under construction. No quality control is performed.	Each county has its own organization and inclusion/exclusion criteria  50 centers were surveyed.  There is no information found over the total number of cases included in these local "registers".	<b>26 cases</b>
<b><u>Informal Sources and Media</u></b>				
<b>Advertisements in three patient oriented journals</b>			A circulation of around 31.000 copies/issue	<b>9 cases</b>
<b>Informal sources</b>				<b>1 case</b>
<b>Total</b>			<b>3,6 million possible events were included in the screening</b>	<b>384 cases</b>

### Questionnaire

In **Study III** a semi-structured questionnaire covering incidence, management, follow-up and prevention of pedSCI, was administrated by mail/e-mail to experts in participating countries (Table 5).

**TABLE 5.** The context of the questionnaire

<i>Context of the questionnaire</i>
<ul style="list-style-type: none"><li>• Published national incidence or empirically based estimates based on the experience of the respondents</li><li>• Organization of initial care and subsequent rehabilitation</li><li>• Organization of follow-up</li><li>• Prevention programs</li><li>• Professionals with special interest in pedSCI</li><li>• Cooperation between adult SCI care and pedSCI care.</li><li>• General/additional comments</li></ul>

### Interviews

In **Study IV** semi-structured interviews were performed. Three themes regarding care and rehabilitation process were chosen:

1. Factors beneficial to the process.
2. Factors impeding the process.
3. Suggestions for improvement of the process.

Interviews expanded by use of further open-ended questions, e.g.: *What do you mean by that?; Can you describe more about this?; What did you think about that?*

## Study populations

Table 6 depicts definitions and criteria used in **Study II**.

**TABLE 6.** The definition, the exclusion criteria, the inclusion criteria and the decision about the codes of diagnosis.

Definition	Exclusion criteria	Inclusion criteria	Requested classification codes from the Swedish National Board of Health and Welfare	
<p>Impairment resulting from traumatic lesions of the cauda equina or the spinal cord proper. Lesions with complete sensory-motor restitution were also included, as defined by American Spinal Injury Association (ASIA), impairment Scale grade E, but with documented evidence of previous symptoms and signs of SCI for at least two weeks after the injury<sup>139</sup>.</p> <p>When the incidence was estimated, mortality directly related to the traumatic event was operationally defined as death occurring within one year post injury<sup>140</sup>.</p>	<p>Cases with isolated root lesions or no injury to the intraspinal neural elements.</p> <p>Cases with congenital malformations of the spinal cord.</p> <p>Cases with acquired SCI of non-traumatic etiology.</p>	<p>The injury must have occurred in Sweden between the years 1-1-1985 until 31-12-1996.</p> <p>At the time of the injury the child should be between 0-15 years of age.</p>	<p>The Swedish version of the 9<sup>th</sup> edition of the International Classification of Disease (rev. ICD 9 used in the period 1987-1995 and their equivalents in the 8<sup>th</sup> edition of this classification)<sup>141,142</sup>.</p>	<p>806 A-X    <i>Fracture of the spine with a SCI</i></p> <p>952 A-X    <i>SCI without any signs of damage to the spinal vertebrae</i></p> <p>767E        <i>Birth injuries, injuries to the spine and the spinal cord</i></p> <p>344 A-X    <i>Other paralyses</i></p>

The study population included three groups of children and adolescents:

1. For cases dead prior to hospital admission (N=43), data from the Cause of Death register<sup>136,137</sup> were used as verification. Autopsy protocols were not reviewed.
2. For cases dead within one year post-injury (N=12), the head of the department at the treating hospital verified the diagnosis by retrospectively reviewing the medical records. The information originated from the Hospital Discharge register<sup>135</sup> and the Cause of Death register<sup>136,137</sup>.

3. For survivors (N=37), retrieved data were checked against the medical records, by requesting such retrospective verification from a senior staff physician at the treating hospital. Subjects were then directly contacted for further verification of diagnosis, after which the remaining putatively true cases were interviewed and in some cases also examined for final verification and additional data retrieval. The information originated from the Hospital Discharge register (N=34)<sup>135</sup>, Habilitation Centers (N=2) and informal contacts (N=1).

The pedSCI population was assessed according to the following descriptors: gender, age at injury, cause of injury<sup>141,142</sup> and cause of death<sup>136,137</sup>.

*Gross neurological and functional outcome* was assessed among survivors by review of medical records and/or additional clinical observations.

*Mortality* directly related to the SCI was operationally defined as all deaths which had occurred within one year post-injury. Subjects that died later than one year post-injury were allocated to the survivor group. (In **Study II**, this comprised two cases who died three years and nine years post-injury respectively).

*Etiological classification:* Through the interviews (**Study II**), ten cases initially classified as fall accidents, and two cases initially classified as traffic accidents, were re-classified as sports injuries.

*Care providers:* The survivors were treated at county hospitals, and/or university clinics with or without specialized SCI units. The data sources utilized were the Hospital Discharge register (HDR), medical records and the patients themselves. If a child had been treated in two or more hospitals, the hospital where the child had been treated the longest period was indicated.

In **Study III** the study population included persons working with SCI in 19 countries. The member states of the European Union (EU) at the time of the study (N=15) were chosen for inclusion, also including Norway and Iceland due to the *EEA (European Economic Area)* agreement<sup>143</sup>. Switzerland and two Baltic countries (Latvia and Lithuania) were also included, due to previous contacts and/or a known interest in pedSCI. Initially the International Spinal Cord Society (ISCoS) membership list was used to identify contact persons. No contact person was identified in Luxemburg why this country had to be excluded from the study.

In **Study IV** the study population comprised a subset of 28 persons. These were the persons who had been adolescents, aged 11-15 years, at time of injury between 1985 and 1996. This subgroup originated from a group of 37 persons aged 0-15 years that had survived at least two years after a SCI and who had been identified by **Study II**.

Out of the 28 persons, one male was excluded due to severe mental retardation, one male had died, and two males declined participation. Thus, 24 persons were included in this study.

Nine had a complete lesion <sup>52</sup>. Fifteen of 24 persons were wheelchair users. Further descriptors are shown in Table 7.

**TABLE 7.** Interviewee descriptors by age at injury, age at interview, gender, level of injury, etiology and hospitalization.

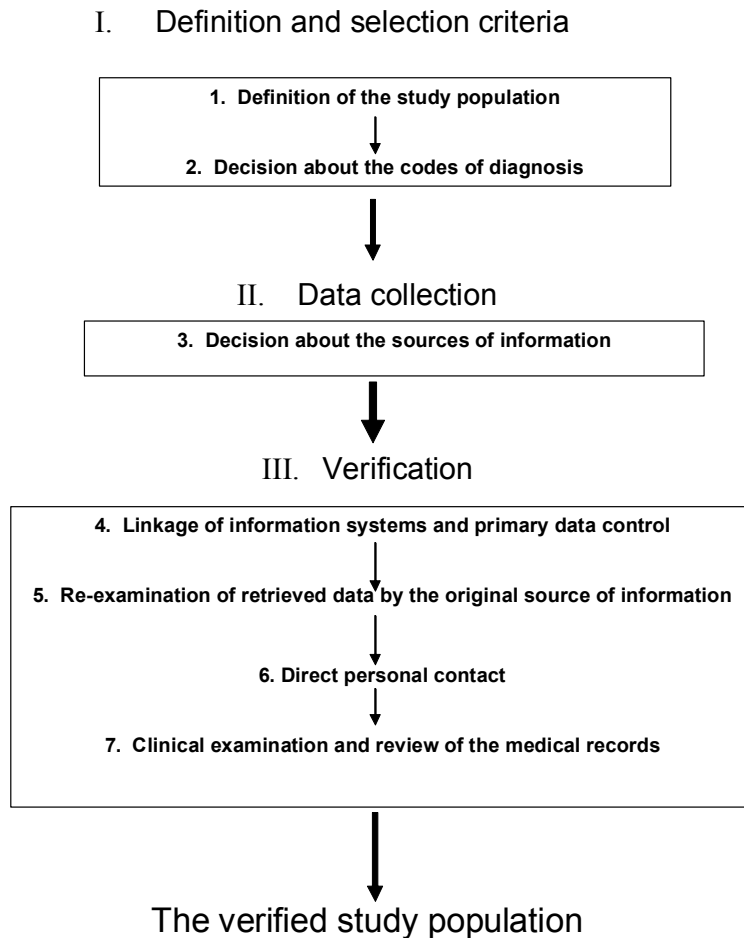
<i>Descriptors</i>	<i>n =24</i>
<b>Age at injury</b>	
Average age, total group	14.1 (SD 4.0, range 11-15 years)
Male	14.5 (SD 0.5, range 14-15 years)
Female	13.5 (SD 1.1, range 11-15 years)
<b>Age at interview</b>	
Average age	24.2 (SD 4.0, range 17-30 years)
<b>Gender</b>	
Male	14
Female	10
<b>Level of Injury</b>	
Tetraplegia	9
Paraplegia	15
<b>Etiology</b>	
Traffic	10
Falls	1
Sports	11
Surgery	2
<b>Hospitalization</b>	
SCI unit	19
Adult orthopedic department	3
Pediatric department	2



## Methods and procedures

In **Study I and II** persons who, at the age of 0-15 years, hade sustained a SCI in Sweden, were identified through population-based registers, local registers and informal sources, as shown in Figure 1.

**FIGURE 1.** Process chart of finding a verified study population



First, the study population was defined by criteria for inclusion and exclusion. A request for data was made, including all ICD codes that conceivably might represent cases comprising the target population.

Verification was made in four steps (Figure 1): 1. Comparisons of data from available sources, in order to avoid duplicates, and for exclusion of false positives. 2. Requests to the original data source, e.g. the treating hospital, for double-checking the correctness of diagnosis. 3. Direct contact with subjects to verify diagnosis and other data from the

medical records. 4. If ambiguity as regards diagnosis and/ or outcome remained, clinical examination and/or additional review of medical records was made.

In **Study III** encompassed contact persons from 19 European countries, i.e. Austria, Belgium, Denmark, Finland, France, Germany, Greece, Iceland, Ireland, Italy, Latvia, Lithuania, the Netherlands, Norway, Portugal, Spain, Sweden, Switzerland and the United Kingdom.

The ISCoS membership list was used to identify professionals with a special interest in pedSCI and/or who had done research on SCI epidemiology. In the absence of such a professional in a given country, representative of a major rehabilitation center was contacted. This person, in turn, was asked to refer the questionnaire to a colleague if such a person was thought to be more knowledgeable as regards the subject matter of the study.

Several reminders were made by phone, email and letters as needed. Questionnaires were sent out in November 2002, and the last questionnaire was analyzed in March 2004, when 18 out of 19 questionnaires had been returned.

In March 2004, a second survey was made. The results of the first survey were summarized in tables and sent to all responders (N=18) for validation. One additional question was included, regarding cooperation between pediatric and adult SCI rehabilitation facilities. Seventeen out of 18 individuals responded to the second questionnaire.

In **Study IV** individual narratives were obtained by semi-structured open-ended personal interviews. Qualitative research is exploratory and inductive by nature. This approach allows more in-depth and comprehensive information. Respondents express their thoughts and experiences in their own words, rather than by predefined response alternatives<sup>134</sup>. The “experienced adolescent” has been found to be imaginative in suggesting ideas and solutions, many of which may not be obvious to health care professionals<sup>88</sup>.

The interviewees were contacted by phone and informed consent for a subsequent interview was obtained. Interviews lasted 60-90 minutes, and were conducted by one of the authors (MA), typically in the home of the interviewee. The recorded interviews were transcribed verbatim. Interviews were performed during 2002-2004, an average of 10 years (4-15 years) after injury.

Narratives were analyzed according to content analysis, i.e. according to a well-established method of identifying, coding, and categorizing patterns in qualitative data<sup>134</sup>

## Analysis and validation

### ***Calculation of estimated incidence- Study II***

The incidence estimates of pedSCI were obtained by calculations based on data from population registers and County Habilitation Centers for the years 1-1-1985 to 31-12-1996.

The total population of Swedish children below 16 years of age during the 12 year period was 20.171.823 children, with an average population of 1.680.000 children/year (max/min: 1.767.000/1.613.000)<sup>144</sup>.

Rather than using the total population to estimate the incidence for the total period, information from each year was used. The incidence for each year was treated as a random variable over the 12-year period. The expected value and the confidence interval were then based on the annual incidence.

### ***Calculation of estimated incidence- Study III***

The midyear population (0-14 years), for each country, was calculated from a publicly accessible database<sup>145</sup> (Figure 2). For the countries who submitted the approximate number of pedSCI cases annually and where the corresponding number of cases at risk was available, a calculation of estimated incidence was made (Figure 3):

$$\frac{\text{Reported occurrence per year}}{\frac{\text{Midyear population (0 - 14 years)}}{15}} \times \text{Number of reported age groups}$$

**FIGURE 2.** Calculation of estimated incidence

The results of the first survey were presented in tables and sent to all responders to the first survey (N=18) for validation.

### **Content analysis –Study IV**

Transcripts in **Study IV** were analyzed by content analysis <sup>134</sup>. This method comprises a stepwise process where “subcategories”, “categories”, and “core categories” are identified inductively. It aims at revealing discrete themes within data. Repetitive themes, or regularities, allow for systematization and classification <sup>134</sup>. Analysis was made in five steps <sup>134</sup>:

- All tapes and transcripts were listened to and read several times for an overall understanding <sup>146</sup>.
- A preliminary coding of each transcript was made, highlighting sentences that captured key concepts.
- The text was read through several times and so-called meaning units were identified. Each such unit comprises words, sentences or paragraphs that cover aspects that are connected through content and context <sup>147</sup>.
- Units were grouped into thematic subcategories. Subcategories were then further aggregated into categories and core categories <sup>134</sup>.
- The process was repeated until no new patterns or themes emerged.

In order to assess concordance within and between narratives, frequency counts of the categorical aggregates were obtained.

Categories were validated by a consensus strategy, whereby the authors discussed and re-examined coding discrepancies in order to reach consistency.

### **Ethical considerations**

The studies were approved by the regional Ethics Committee (permit no. Um dnr 96-289) and by the Swedish Data Inspection Board.

In **Study I**, informed consent was obtained once subjects were identified and verified (Figure 1, step 5).

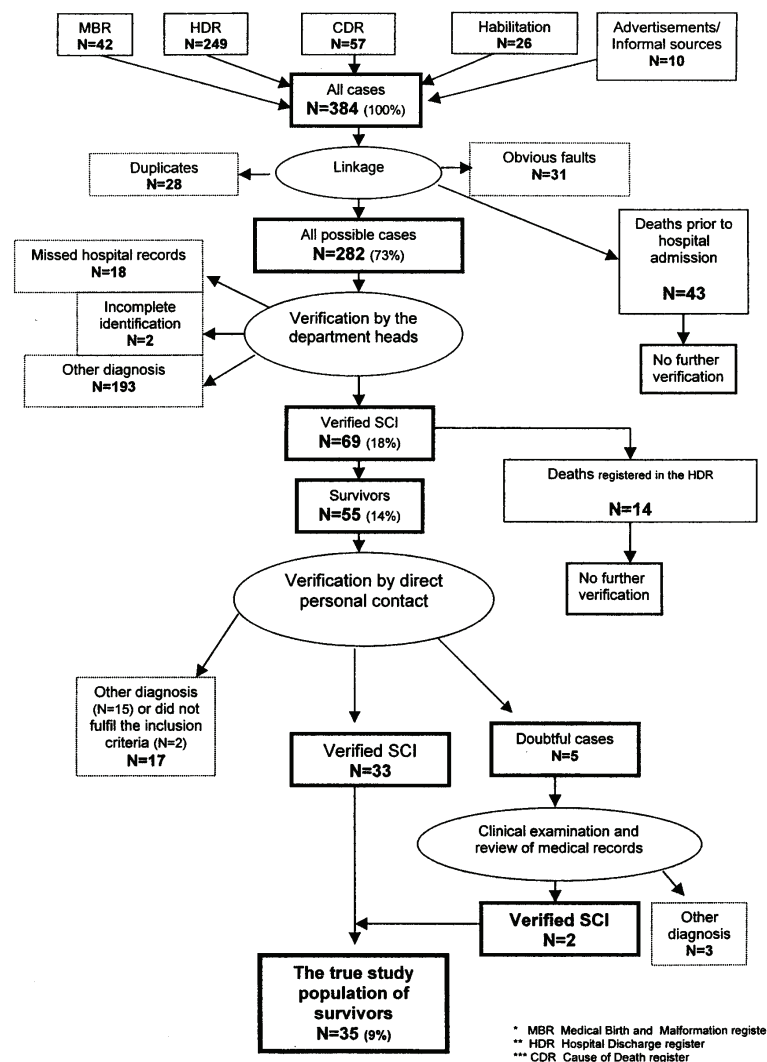
In **Study IV** the interviewees gave informed consent and were granted confidentiality. Individual interviewees were guaranteed not to be identifiable in the subsequent report. Interviewees were informed that they could terminate their participation in the study at any time. The interviewer (MA) was not involved in the medical care of any interviewee at any time.

## MAIN RESULTS

- *How to find and verify pediatric SCI in Sweden?*

### Study I

The registers of the Swedish National Board of Health and Welfare, Habilitation Centers and various “informal” sources were all used for identification of the study population.



**FIGURE 3.** Flow-chart of the process of finding children and adolescents (0-15 years) with pedSCI in Sweden, 1985-1996.

Initial screening identified 384 cases, which by subsequent analysis were found to include a large number of false positives (i.e. incorrect diagnostic coding). Ultimately, 35 living cases could be fully verified and 14 deceased cases could be pragmatically verified (Figure 3).

Verification of cases identified by initial screening was made by (1) linkage of information from the various data sources (Table 8), (2) by a senior staff physician at the treating hospital (Table 9) and (3) by direct contact with the cases (Figure 3).

**TABLE 8.** Linkage of information systems and primary data control.

<b>All cases N=384</b>			
<i>Included</i>		<i>Excluded</i>	
<b>Further verification</b>	<b>Obvious faults</b> N= Excluded cases according to the set definition and the inclusion criteria in different sources.	<b>Duplicates</b> N= Cases registered in the HDR and in an additional source.	<b>Deaths prior to hospital admission</b> N= cases found exclusively in the CDR.
282 children identified with a possible SCI	HDR N=12/249 Habilitation N=16/26 Advertisements N=3/9	CDR N=14/57 Habilitation N=8/26 Advertisements N=6/9	Of the 57 children with suspected SCI identified through the CDR, 43 children had died prior to hospital admission and subsequently not found in the HDR. In the example, no further reviews of their autopsy protocols were obtained.
<b>282 cases</b>	<b>31 cases</b>	<b>28 cases</b>	<b>43 cases</b>

**TABLE 9.** Description of the requested codes of diagnosis in the Hospital Discharge Register (HDR), the Medical Birth and Malformation register (MBR), cases from the Habilitation Centers, informal sources and the result after verification by the department heads.

<i>Sources and Classification codes (rev.ICD 9)</i>	<i>Total checked</i>	<i>Other diagnosis</i>	<i>Missing records</i>	<i>Incomplete identification</i>	<b><i>“True” SCI</i></b>
<b>HDR</b>					
<i>SCI with fracture 806</i>	211	144*	9	0	<b>58</b>
<i>SCI without fracture 952</i>	21	14	1	0	<b>6</b>
<i>Birth injury 767E</i>	3	1	1	0	<b>1</b>
<i>Other paralysis 344</i>	2	1	0	0	<b>1</b>
<b>MBR</b>					
<i>Birth injury 767E</i>	42	33	7	2	<b>0</b>
<b>Habilitation centers</b>	2	0	0	0	<b>2</b>
<i>Cases reported as having SCI</i>					
<b>Informal sources</b>	1	0	0	0	<b>1</b>
<i>Cases reported as having SCI</i>					
<b>TOTAL</b>	<b>282</b>	<b>193</b>	<b>18</b>	<b>2</b>	<b>69</b>

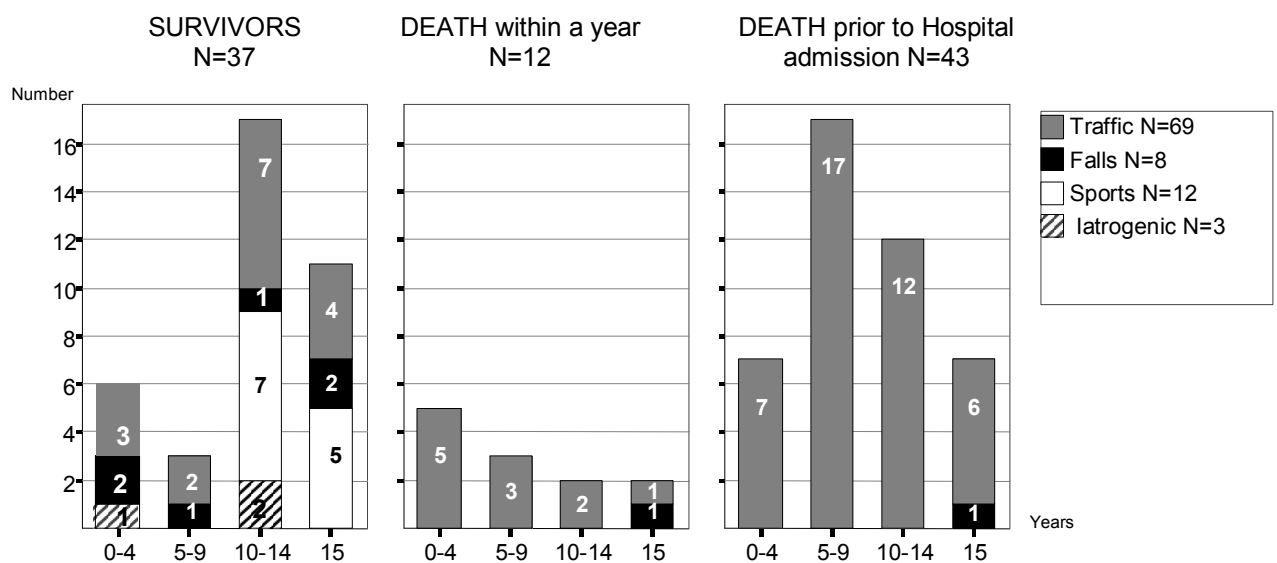
\*128 cases were verified by the department heads as having sustained code 805, *Fracture of the spine without a SCI*.

This study showed that cases registered by ICD-codes 806 (*SCI with fracture of the spine*) and 952 (*SCI without fracture of the spine*) included over 70% “false positives”, i.e. less than 30% “true” SCI (Table 9). Quality control was found to be time consuming but necessary in order to verify true cases.

- *How frequent is pediatric SCI in Sweden?*
  - *How many have survived the injury?*
    - *What were the causes?*
  - *Where were the children treated?*

## Study II

The incidence was 4.6 /million children/year (95% CI 3.6-5.5). When excluding pre hospital fatalities, the incidence was 2.4 (95% CI 1.8-3.1).

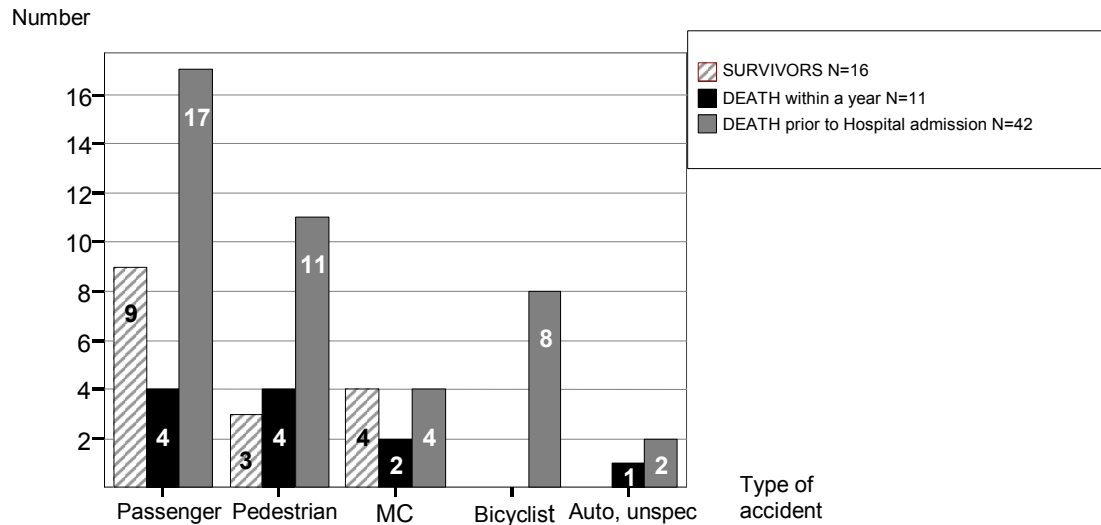


**FIGURE 4.** Causes of injury by age group and outcome, N=92.

Associated injuries occurred in 41% (N=38) of the cases. The mortality was high. Fifty-five cases (60%) of the total group (N=92) died within one year after injury.

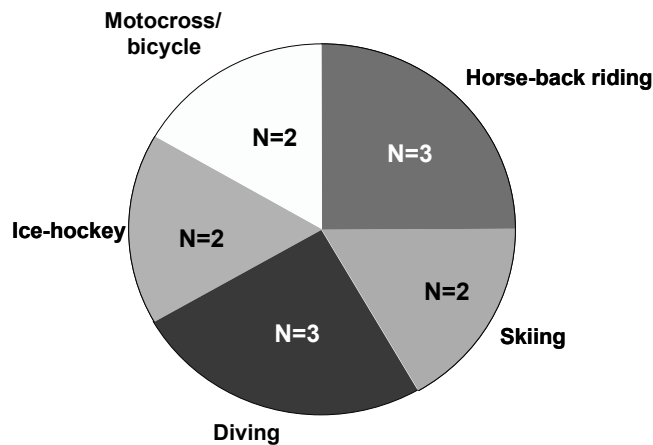
The main cause of injury among fatalities was traffic accidents (Figure 4). The mean age was 9.2 years (SD 3.6; range 4-15). The pedestrians had a mean age of 8.9 years (SD 2.8; range 5-14) (Figure 5)





**FIGURE 5.** The distribution of traffic accidents

The second leading cause was sports related injuries. Among survivors (10-15 years), sports related injuries (43%) were as common a cause as traffic accidents (39%) (Figure 4). The causes are shown in Figure 6.



**FIGURE 6.** Distribution of sport-related injuries among adolescents 13-15 years.

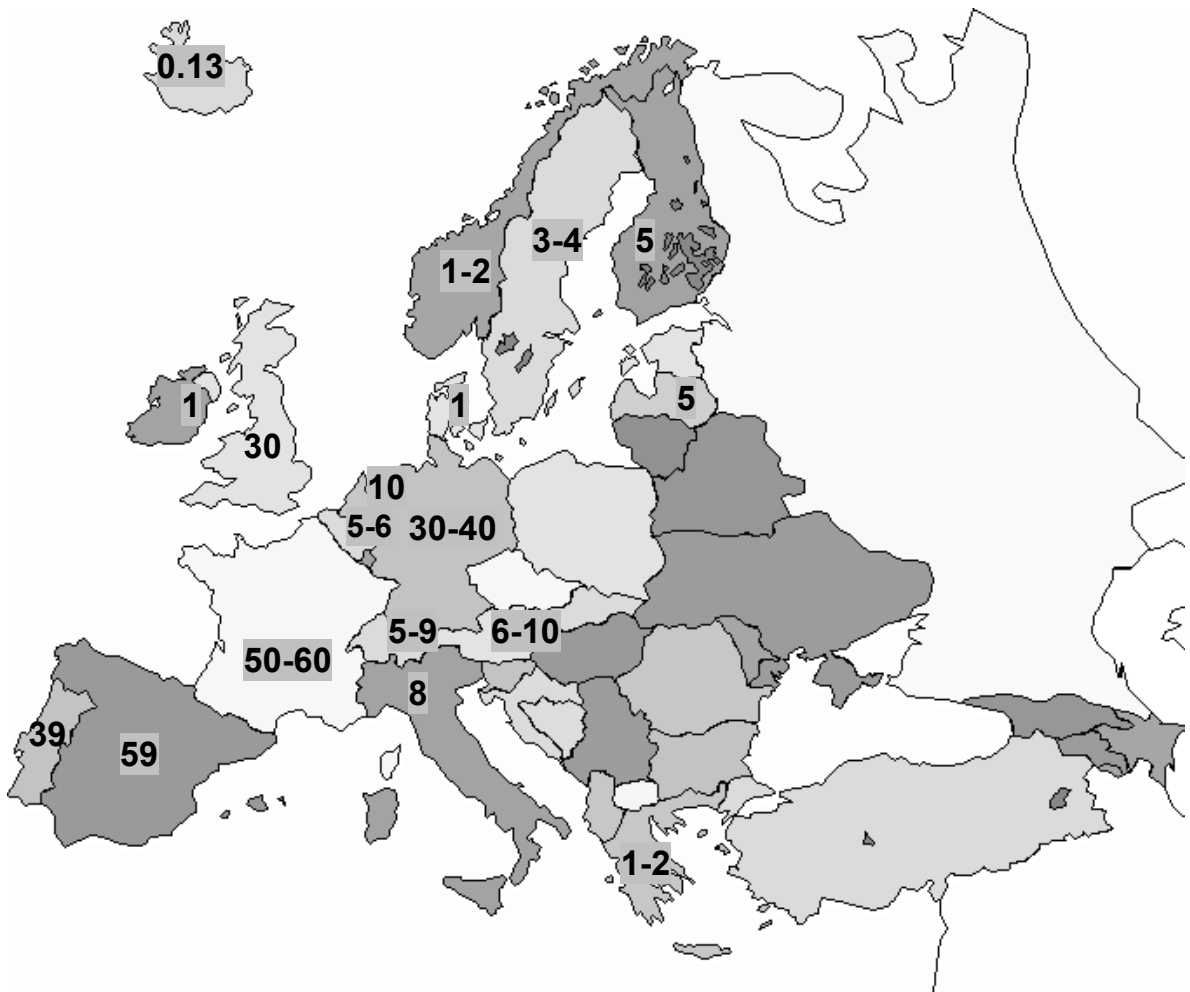
The survivors were treated in 18 different hospitals.

The average age of cases treated at an SCI unit was 14 years (SD 1.3; range 9-15 years), whereas cases treated elsewhere had a lower average age, 8 and 8.9 years, respectively.

- *How frequent is pediatric SCI in other countries in Europe?*
  - *Where are they treated?*
  - *Is there any interest for cooperation in treatment and research?*
- 

### Study III

The reported numbers of cases of pedSCI per year are shown in Figure. 7.



**FIGURE 7.** Reported estimated number of pedSCI/year from 18 countries.

Only in Portugal and Sweden, is the incidence of pedSCI established with some degree of certainty, and was 27 cases/million children/year and 4.6 cases/million children/year respectively (fatalities included). For all countries, the estimated incidence of pedSCI, non-fatal injuries, varied from 0.9-23 cases/millionchildren/year in the ages 0-14 years (Table 10).

Thus, although the incidence varies considerably, it is concluded that pedSCI is rare throughout Europe (Table 10).

The total annual number of pedSCI in 18 European countries based on these rough estimates was thus less than 300 cases (Figure 7). The midyear population of children aged 0-14 years/million in this region was 64.642.<sup>145</sup>

If extrapolating from the figures of a “low incidence country” such as Sweden, there would be a rough estimate of 155 cases surviving pedSCI each year in the 18 surveyed European countries. If extrapolating from the figures of a “high incidence country” such as Portugal there would be a rough estimate of 1466 cases annually. An average of these extremes yield the estimate of just over 800 children in the ages 0-14 years surviving with a SCI in this region.

Initial care and follow-up differ between the countries depending on the age of the child and the local organization of health care (Table 11,12). Several countries have programs for pediatric injury prevention and road safety legislation e.g. related to seatbelts, car seats, helmets, and speed limits. Available prevention programs specifically aimed at SCI are mainly focusing on young adults (Table 10).

It is concluded that most countries lack reliable statistics concerning pedSCI. Many respondents (N=14) sought more collaboration in this field, in order to exchange knowledge in research and to further develop clinical practice guidelines.

**TABLE 10.** Incidence of pedSCI, estimated number of new cases with SCI/year, age, midyear population (0-14 years)/million <sup>145</sup>, estimated incidence and pedSCI prevention efforts in 18 European countries.

Country	Incidence of pedSCI. Cases/ million children/year)	Estimated number of new cases with SCI/year	Age (years)	Midyear population by (0-14 years)/million <sup>145</sup>	Estimated incidence <sup>1</sup> .	PedSCI Prevention/ comments
Austria		6-10	3-10	1.316	8.5-14.2	No/General trauma
Belgium		5-6	5-15	1.777	3.8-4.6	No
Denmark		1	0-15	1.015	0.9	--/Road Safety- Adult SCI
Finland		5	0-16	0.922	4.8	Yes/Adult SCI
France		50-60	0-15	11.175	4.2-5.0	Yes/Road Safety
Germany		30-40	0-15	12.301	2.3-3.0	No
Greece		Less than 10 (in 5 years) ≈2) (Data only from one Neuropathic Bladder Unit)	6-13	1.544	--	No/unsure
Iceland		4 (in 30 years) = 0.13	0-15	0.063	1.9	Yes/General child trauma
Ireland		1	4-17	0.829	1.3	Yes/Road safety
Italy <sup>148</sup>		16 (1997-1999) =5.3	0-15	8.141	--	Unknown/ Adult SCI
Latvia		5	7-16	0.353	21.2	No
Norway <sup>149</sup>	1.5	14 (1991-2001)= 1.4	0-15	0.907	1.4	No/general trauma
Portugal <sup>21</sup>	27*	46*/ 16% reported mortality rate prior to hospital admission =39**	0-14	1.699	27.0*/23**	No
Spain		59	0-14	5.794	10.2	Yes/in schools
Sweden <sup>140</sup>	4.6*/ 2.4**	7*/4** (based on an average population of 1.68 million in the ages 0-15 years/year)	0-15	1.597	4.1*/2.4**	No/Road safety and general trauma
Switzerland- Swiss Paraplegic Center		5-9 (+ 40%) ≈ 7-13	0-14	1.255	5.6-10.4	No answer
The Netherlands		10 (Data only from the Hoogstraat)	0-20	2.983	--	No
United Kingdom <sup>150</sup>	5.2* /2.8**	60*/30**	0-14	10.971	5.5*/2.7**	Yes/ <a href="http://www.rosipa.com">www.rosipa.com</a>

\*cases dead prior to hospital admission are included

\*\*survivors

<sup>1</sup>

$$\frac{\text{Reported occurrence per year}}{\frac{\text{Midyear population (0 - 14 years)}}{15}} \times \text{Number of reported age groups}$$

**TABLE 11.** Initial care and rehabilitation in different settings of care for children (0-15 years) with SCI in 18 European countries.

Settings of care	Pediatric department at a <b>local</b> hospital (no specialized rehabilitation).			Pediatric department at a <b>regional</b> hospital (no specialized rehabilitation			Specialized pediatric rehabilitation department/ neuropaediatric			Specialized SCI unit for adults.			General rehabilitation department for adults.			Other departments (i.e. university traumatology)		
	0-6 years	7-12 years	13-15 years	0-6 years	7-12 years	13-15 years	0-6 years	7-12 years	13-15 years	0-6 years	7-12 years	13-15 years	0-6 years	7-12 years	13-15 years	0-6 years	7-12 years	13-15 years
Countries																		
Age																		
Austria	1	2	2		2	2	3	3	3	1	1	1	3	3	2			
Belgium							1	1	1									
Denmark							1	1	1	1	1	1						
Finland	3			2	2		1	1	1	3	3	1	3			1	1	1
France	2	2	2	2	2	2	1	1	1	3	3	3	3	3	2	3	3	3
Germany	3	2	3	2	2	2	1	1	1	2	2	2	3	3	3			
Greece	2	2	3	1	1	2	3	3	3	3	2	1	3	2	1		3	3
Iceland	3	3	3	1	1	1	3	3	3	3	3	3*	3*	3*	3*	3	3	3
Italy	3	3	3	1	1	1	2	1	1	3	2	2	3	3	3	2**	2**	2**
Ireland	First weeks	First weeks	First weeks	First weeks	First weeks	First weeks	1	1	1									
Latvia	3	3	3	1	2	3	1	2	3	2	1	1	3	3	3	2		
The Netherlands	First weeks	First weeks	First weeks	First weeks	First weeks	First weeks	1	1	1	1	1	1	2	2	2			
Norway	2	2	2	2	2	2	1	1	2	1	1	1	3	3	3			
Portugal				1	1	1												2
Spain																		
Sweden	1	1	1	1	1	1	2	2	2	3	2	1	3	2	3			
Switzerland	2	2	2	1	2	2	2	2	2	2	1	1	3	3	2	1	3	3
UK	1	1	2	1	1	1	2	2	2	2	2	1	3	3	3			

**1 = Often**, 2 = Seldom, 3 = Never \* In cooperation with a specialized SCI team \*\*treated outside the country

**TABLE 12.** Follow-up in different settings of care for children (0-15 years) with SCI in 18 European countries.

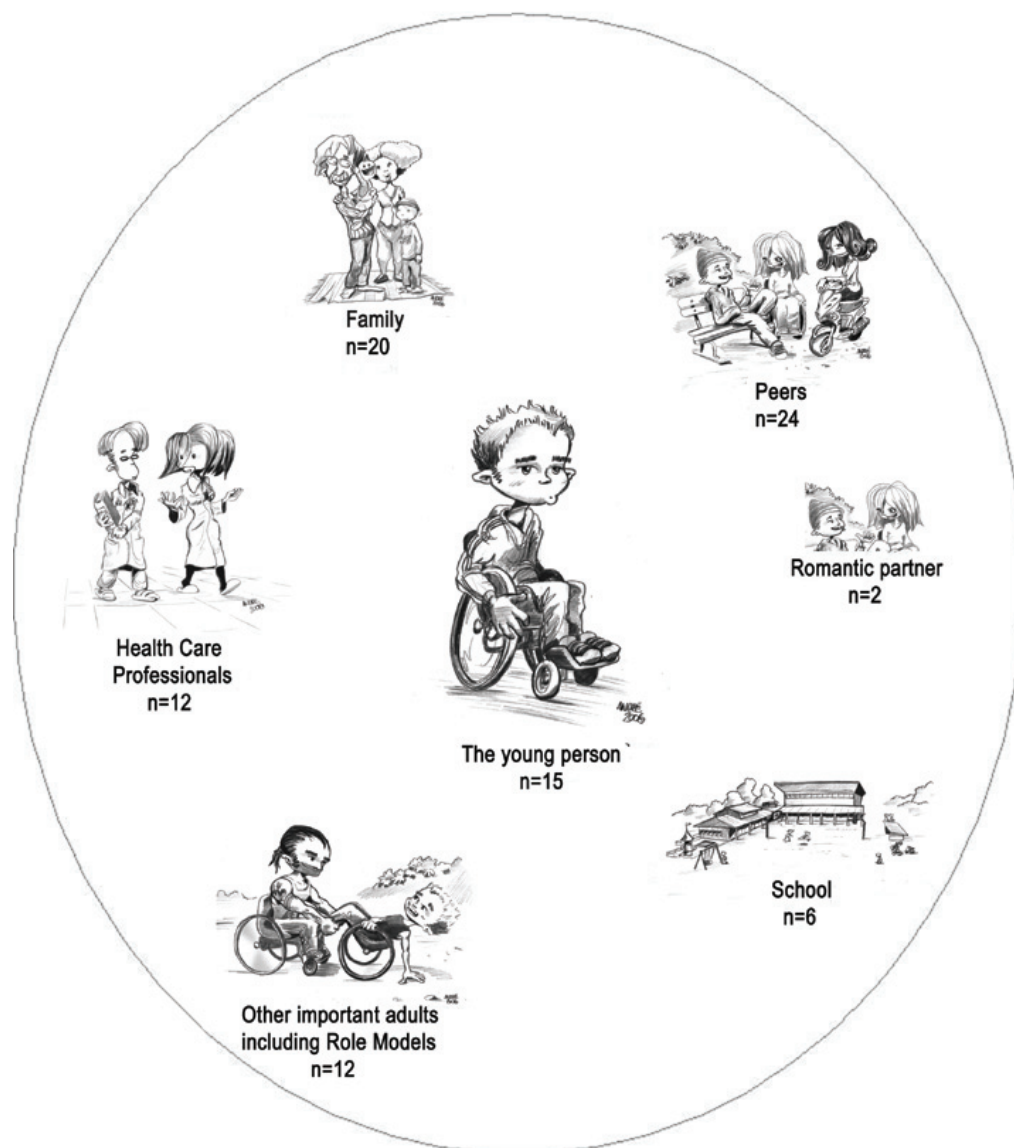
Settings of care	Pediatric department at a local hospital (no specialized rehabilitation).			Pediatric department at a regional hospital (no specialized rehabilitation)			Specialized pediatric rehabilitation department/ neuropaediatric			Specialized SCI unit for adults.			General rehabilitation department for adults.			Other department: Out-patient pediatric rehabilitation/Habilitation		
	0-6 years	7-12 years	13-15 years	0-6 years	7-12 years	13-15 years	0-6 years	7-12 years	13-15 years	0-6 years	7-12 years	13-15 years	0-6 years	7-12 years	13-15 years	0-6 years	7-12 years	13-15 years
Countries																		
Age																		
Austria																		
Belgium							1	1	1									
Denmark							1	1	1			1						
Finland					1	1	1	1	1			1						
France	2	2	2	2	2	2	1	1	1	3	2	2	3	2	2	3	3	3
Germany	2	2	2	2	2	2	1	1	1	2	2	1	3	3				
Greece				1	1	1										1	1	1
Iceland	3	3	3	1	1	1	3	3	3	3	3	3	1*	1*	1*			
Italy	2	3	3	1	2	2	1	1	1	2	2	2	2	3	2	2**	2**	2**
Ireland							1	1	1									
Latvia	3	3	3	2	2	2	1	2	2	2	1	1	3	3	3	1	1	1
The Netherlands	3	3	3	3	3	3	1	1	1	1	1	1	2	2	2			
Norway	1	1	1	1	1	1	1	1	1	1	1	1	3	3	3			
Portugal							1	1	1						2			1
Spain																		
Sweden	1	1	1	1	1	2	2	2	2	2	2	1	3	3	2	1	1	2
Switzerland	2	2	2	1	2	2	2	2	2	2	1	1	3	2	2			
UK	1	2	2	1	1	1	2	2	2	2	2	2	3	3	3			

**1 = Often**, 2 = Seldom, 3 = Never \* In cooperation with a specialized SCI team \*\*treated outside the country

- *What and who did persons who sustained a SCI during adolescence recall as having facilitated or impeded their rehabilitation process?*

#### Study IV

A number of psychosocial factors that were reported as having influenced the rehabilitation process and outcome could be identified. These factors corresponded to the actions of certain key agent(s), see Figure 8.



**FIGURE 8.** Frequencies of key agents (core categories), recalled by the interviewees as having promoted the rehabilitation and early re-adjustment after SCI, n=24.

Specific actions within these core categories comprised a number of psychosocially relevant subcategories. These are summarized in Table 13.



### *The young person*

Having a “fighting spirit” was frequently (15/24) expressed as a facilitating factor. Outwardly directed anger and aggression was common. Frustrations directly related to the presence of physical disability were frequently recalled, but also comments reflecting resilience.



### *The family*

Parents played key roles in the rehabilitation process, e.g. as spokespeople and advocates. They were typically trusted supporters as discussion partners and “pep-talkers”. Parents “absorbed” sorrow, frustration and anger that interviewees hesitated to share with staff.

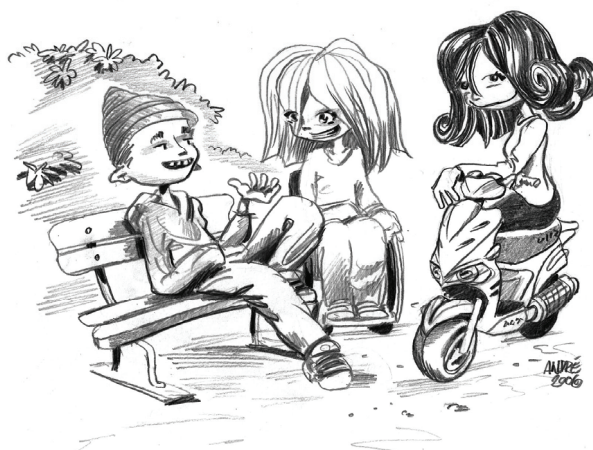




*"I think I turned a lot of this anger and frustration against my parents..."*

Frequently, hospital staff was perceived to having been insensitive to the important role played by the parents. The role of parents as caregivers, however, was seemingly "double-edged", especially regarding intimate body functions, as expressed in the following examples:

*"Yeah, it feels really stupid, it felt really stupid with my mom helping me shower and stuff. But Mom told me that I shouldn't find it embarrassing because she did it when I was a baby/ / But it was really embarrassing, actually".*



### **Peers**

All (24/24) interviewees indicated that peers and friends had played a key role.

*"Many times, I felt that I just wanted to stay at home, but they (the friends) more or less carried and dragged me and the wheelchair out of the house. This, to have my friends around, was probably what made me come back again. They didn't let me sit at home; they basically forced me to get moving..."*

Many interviewees apparently lost contact with their old circle of friends, but managed to find a new social context.



### *Romantic partner*

Only three interviewees (one male and two females) had a romantic partner at the time of the injury. Two of these described that their boyfriends had been very important for emotional support and intimacy, even if time together was restricted, e.g. by visiting hours.



### *Other significant adults*

Twelve of 24 interviewees indicated that persons outside their core family had acted as support and provided pep-talk. These were friends of the family, coaches, or health care professionals who they continued meeting after discharge from hospital.

The importance of a role model was sometimes seemingly great. Compatible age and disability between coach/"role model" and oneself were recalled by some interviewees as being of importance.



### ***School network***

The social network from school, e.g. teachers and classmates, were indicated by 6 of 24 interviewees as having been of importance, and return to school was recollected as an important milestone by some interviewees. The presence of this category emphasizes the importance of staying in touch with teachers and classmates.



### ***Health care professionals***

Health care professionals played an important role as facilitating the psychological aspects of rehabilitation as indicated by 12 of 24 interviewees. Staff was typically appreciated for being knowledgeable, concrete, structured, and empathetic. However, there were also frequent reports of clashes between staff and patients.

**TABLE 13.** Core categories, categories and subcategories facilitating or impeding rehabilitation and early re-adjustment after SCI.

Core category	Category	Subcategories
The young person	Personality characteristics	<b>FACILITATORS</b> Energy/Will Optimism Self confidence <b>IMPEDERS</b> Shame Isolation Anger Anxiety, fear – distrust of staff Docility Obsessive-compulsive behavior (e.g. eating disorders, obsession with medical procedures)
	Growth and pubertal development	<b>IMPEDER</b> Negative appreciation of appearance
	Impact of injury	<b>FACILITATOR</b> Experience of emotional growth <b>IMPEDER</b> Physical impairments regarding mobility, bodily functions, appearance, sexuality, identity, integrity
Family	Parents	<b>FACILITATORS</b> Spokespeople; advocates for the adolescents Pep talk – moral support "Punching bags" <b>IMPEDERS</b> Parents as caregiver Violating integrity Over-involvement
	Siblings	<b>FACILITATORS</b> Pep talk – moral support Older sibling discussing sexuality <b>IMPEDER</b> Sibling as caregiver
Peers		<b>FACILITATORS</b> Connectedness and continuity Identity promotion Activity promotion <b>IMPEDERS</b> Prejudices Non-involvement Long hospitalization, long distances from home, awkward visiting hours Lack of knowledge and understanding
Romantic partner*		<b>FACILITATOR</b> Intimacy <b>IMPEDER</b> Shame Isolation Breaking up
Other significant adults		<b>FACILITATORS</b> Pep talk – moral support Role models for identity development
School	Teachers	<b>FACILITATORS</b> Involvement Connectedness and continuity Identity promotion <b>IMPEDERS</b> Long hospitalization Lack of knowledge
	Classmates	<b>IMPEDERS</b> Long hospitalization Lack of knowledge

**TABLE 13 (continued).** Core categories, categories and subcategories facilitating or impeding rehabilitation and early re-adjustment after SCI.

Core category	Category	Subcategories
Health care professionals	Knowledge	<b>FACILITATORS</b> Biopsychosocial development in adolescence Age-appropriate demands and challenges Adolescents' sexual development Family interaction during adolescence Impact of SCI on adolescent and on family function Importance of peers and significant others
	Attitudes	<b>FACILITATORS</b> Being empathic without being intrusive Respecting integrity, and autonomy of the patient Respecting adolescents' sensitivity about naked body and bodily functions Providing flexibility and extra time when needed Respecting parents expertise and parenting style Recognizing peers and romantic partners as instrumental for age-appropriate development <b>IMPEDERS</b> Feeling sorry for the patient Seeing parents as a burden rather than as a resource Excluding the family
	Skills	<b>FACILITATORS</b> Providing concrete discussions with checklists on relevant tasks Being able to discuss sexuality in an age-appropriate way Providing age-appropriate recreational activities, if possible with age-mates Balancing independence versus assistance Providing family support Involving the family in an age-appropriate way Providing schooling in hospital with individual tutoring Facilitating school re-entry process <b>IMPEDERS</b> Talking with parents over the patient's head Inability to cope with emotional reactions of the patient Not involving the patient in age-appropriate decisions, i.e. food choices, visiting hours Working according to protocols without concern for patients' actual needs

## DISCUSSION

Four issues related to this thesis will be briefly discussed:

- Improved data collection and research design in the epidemiology of pedSCI.
- Development of prevention programs.
- High quality care and research despite the rarity of the disorder.
- Implementation of insights from adolescence medicine.

### *Improved data collection and research design in the epidemiology of pedSCI*

In contrast to most other countries, Sweden has several readily available population based registers. These registers have very low drop out rates, and misdiagnosis in general does not exceed about 10 percent <sup>137</sup>. Unfortunately, these registers seem unreliable when applied to pedSCI. The frequency of false positive diagnoses found in **Study I** renders the HDR virtually useless for direct analysis. Additional quality control is necessary. Furthermore, in order to allow meaningful meta-analysis, the set definition and inclusion/exclusion criteria have to be specified in detail in order to avoid incompatibilities between studies. Of particular importance is the setting of the upper age limit for what is defined as pedSCI. A case in point is a recent article from the US <sup>34</sup>, reporting an incidence of “pediatric” SCI of 20 children/million children/year. The incidence population includes cases aged 0-18 years, and the study design does not provide any further separation into age groups. Such pooling brings together completely disparate cases, e.g. a toddler being a passenger in a car and an 18-year old biker high on cocaine. The epidemiological study of pedSCI is likely to benefit from developmentally based age-at-injury groupings, thereby considering the varying psychological and behavioral characteristics from infancy through adolescence <sup>19</sup>.

In order to increase comprehensiveness, all diagnostic classification codes (ICD) that might reasonably be used to represent the set definition should be considered for inclusion in the initial screening. Unfortunately, classification codes are not universally comparable <sup>141,142,151</sup>.

Furthermore, different revisions of a specific diagnostic manual may lack comparability; making it difficult to pinpoint which diagnostic code(s) that may have been used to represent pedSCI. As can be seen in **Study I**, this was the case even with the restricted purpose of identifying ped SCI of a *traumatic* etiology. The identification of spinal cord lesions of *non-traumatic* etiology in the HDR would have been next to impossible according to our opinion.

One possible solution to these problems is the development of specific “quality registers” for rare diagnoses such as SCI. Several such registers have in fact been developed by the National Board of Health and Welfare in Sweden. The SCI Council in Sweden has produced a consensus document on basic quality indicators of SCI care, as well as a set of descriptors of treatments and outcomes. A national database which collects data from all major national SCI units was launched in 1997. This data base is a potentially useful source for research projects and may facilitate collaborative projects <sup>124,152</sup>. Internationally, the development of a so-called core data set has further standardized and improved the collection and reporting of SCI <sup>13,14</sup>. It is obviously of importance to also determine the reliability of these standards for pedSCI <sup>53</sup>.

Despite these efforts, there remains the problem of altogether missed cases in available registers. The care of pedSCI in Sweden remains severely scattered in a non-systematic way, well beyond the SCI centers, as indicated by **Study II**. Especially the youngest children are treated in pediatric settings which lack contact with adult SCI centers and which are unaware of the national SCI database and/or lack familiarity with the SCI core data set.

### ***Development of SCI prevention programs***

Traffic accidents are the main cause of pedSCI, as found in **Study II**. The incidence of pediatric trauma in Sweden is among the lowest in the world. This is likely a result of vigorous preventative work performed over the last 50 years. Laws and regulations have made the environment for the child safer, as have systematic information and education efforts aimed at their parents <sup>153,154</sup>. Still, accidents remain the most common causes of death among children and adolescents aged 1-14 years in Sweden <sup>153,154</sup>. Children are vulnerable in traffic due to the complex situations that frequently arises and which exceed their cognitive, developmental, behavioral, physical and sensory abilities. Children are impulsive and have difficulties in judging speed, spatial relations and distances. Additionally, adolescents in particular tend to be risk-prone and easily influenced by emotions, stress, and peer pressure <sup>155</sup>. They frequently act in the adult environment, but they lack the experience and mature behavior of an adult.

Sport is another leading cause of pedSCI in Sweden , especially so among adolescents, This is in accordance with reports from other countries <sup>36,38,39,156,157</sup>. Preventative measures, e.g. strict rules and adequate equipment, should be ensured in sports carrying a risk for SCI.

SCI prevention should be tailored according to the specific risk profile of each age group. As Sweden until now has focused most prevention strategies on younger children <sup>132</sup>, there is now a need for national coordination and methodological development of preventative efforts towards adolescents.

***Provision of high quality care and research despite the rarity of the disorder***

The low incidence and prevalence of SCI in Sweden (**Study I**) and in many countries in Europe (**Study III**) may underlie the unsystematized care and low priority given to development of research, care and prevention programs within the field of pedSCI.

SCI is physiologically complex and affects many organ systems<sup>54</sup>, and therefore requires the input from many organ specialists. It is well-documented that comprehensive management and systematic follow-up will improve outcomes<sup>16,121</sup>. One key feature of the Model SCI systems is sufficient volumes of patients, something which necessitates some degree of centralization<sup>16</sup>. This certainly holds true for the even rarer event of pedSCI.

The low incidence of SCI makes epidemiological studies problematic<sup>36</sup>. One obvious strategy is pooling study populations by multi-center studies.

An example of a successful international network from another field is the Paediatric Rheumatology International Trials Organisation (PRINTO), supported by the European Economic Community (EEC). PRINTO has grown rapidly and now (2007) includes 47 countries not only from Europe but also Latin America, Africa, Australia and New Zealand and Asia. Its aim is to facilitate and conduct high quality research, and also to provide information to the children and their parents<sup>158</sup>.

Surprisingly, pedSCI it is not currently included in the list of rare diseases provided by The European Organisation for Rare Diseases (EURODIS)<sup>159</sup>. A “rare disease” is defined as a “life-threatening or chronically debilitating disease, which is of such low prevalence that special combined efforts are needed to address it. As a guide, low prevalence is taken as prevalence of less than 5 per 10,000 in the Community”<sup>160</sup>. PedSCI, actually fulfilling this definition, would benefit from being included in this group of diseases.

Thus, there is a need for increased collaboration between countries. The establishment of a European/international pedSCI network, would likely be of benefit to all professionals and, most importantly, to the children with SCI and their families.



### *Implementation of insights from adolescence medicine*

The involvement of the social network of the injured young person, comprising parents, peers and friends, is important for successful rehabilitation. Within pediatrics and adolescence medicine this is well-known, but in adult care settings it may be less recognized. Most persons with pedSCI were treated in adult settings and the health care providers were not typically perceived as promoting involvement of the social network. If a child or adolescent is treated in an adult setting, rehabilitation professionals might be encouraged to increase their knowledge of pediatric and adolescence medicine in order to better meet the specific needs and demands of children in different age groups.

Meeting other persons with a similar disease is regarded as an important care component for young persons with chronic illness<sup>7,90,94,96,113</sup>. As pedSCI is rare, such meetings will not occur spontaneously. How to promote such interactions must be further discussed.

Increased communication between pediatric hospital departments and SCI units is needed, both as regards individual patients as well as regards registration and data collection. To further improve care and research an increased exchange of information and best practice between countries in Scandinavia and Europe is of the essence.

### **Limitations**

First, the incidence estimates based on **Study I** and **II** may possibly be too conservative due to the likely omission of the SCI diagnosis in cases of severe fatal multi-trauma, as discussed by Dickman<sup>7</sup>.

Secondly, the incidence estimates in **Study III** implied large variations between countries. Countries with a similar socio-economic profile would be expected to have fairly similar incidences, but differences nevertheless were found. This may of course be due to true incidence variations but may also, and perhaps more likely, be due to scattered care, varying reporting methods, whether fatal accidents are included or not and/or due to differences in operational definitions of the diagnosis. Whatever the case, the incidence estimates reported in this thesis have to be considered highly preliminary. Subsequent studies may benefit from utilizing other methods for data collection, e.g. the Delphi method<sup>161</sup>. This method allows structured collection of information in order to reach consensus by a group of geographically dispersed experts<sup>161</sup>.

Apart from the inherent and perhaps inevitable limitations of qualitative methodology, the most obvious limitation of **Study IV** is the recall bias imposed by the years gone by from injury to interviews. However, recollections as such are of significance, regardless of their “objective” validity.

Furthermore, the results reflect experiences consequential to the Swedish health care system of 10-20 years ago. One might question whether these experiences are still of relevance. According to **Study III**<sup>162</sup>, however, that state of the health care system seem to differ little, if any, from what is still typically the case in Sweden and most parts of Europe, today.

### **Implications for future research**

Studies with a longitudinal prospective design are needed to build a more comprehensive knowledge base on pedSCI. Besides core knowledge on SCI care, such studies may focus on how developmental needs are met and whether that may improve the long-term outcome on quality of life for children and young persons with SCI.

## CONCLUSIONS

### *Articles    Conclusions*

- I     Registers offer a convenient source for study population identification. However, screening of specific ICD-codes yielded over 70% “false positive” cases of pedSCI. Thus, further refinement and quality control is necessary in order to ensure validity. Such further verification is time-consuming, but nevertheless necessary in order to identify a true cohort.
- II    PedSCI is rare in Sweden. Mortality is high (60%). The main cause of injury is traffic-related. Incidence rates are higher for adolescents aged 14-15 years. Among adolescents injuries caused by sport is as common as injuries in traffic. Care and rehabilitation is fractioned and non-systematized.
- III   The incidence of pedSCI is low throughout Europe. The reported incidence varies considerably between countries. The systems of care also vary considerably.
- IV   Parents and peers play an important role in rehabilitation of pedSCI. Parents frequently act as advocates in interactions with health care providers, as supporters and as containers of sorrow, frustration and anger. Peers act as promoters of activity and identity development. Health care providers are perceived as not sufficiently facilitating involvement of this network. Rehabilitation of adolescents with SCI might improve by encouraging staff to acquire knowledge in adolescence medicine in order to better understand the specific needs and demands of persons of this age. It is further suggested that the active participation of parents and peers will facilitate the rehabilitation effort of the young person with a SCI.

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