Official Journal of the European Paediatric Neurology Society

Original article

Non-traumatic spinal cord ischaemia in childhood – Clinical manifestation, neuroimaging and outcome

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ARTICLE INFO

Article history:
Received 11 April 2012
Received in revised form
28 August 2012
Accepted 1 September 2012

Keywords:
Spinal cord ischaemia
Anterior spinal artery syndrome
Clinical manifestation
Outcome
Neuroimaging
Children

ABSTRACT

Background: Spinal cord ischaemia is rare in childhood and information on clinical presentation and outcome is scarce.

Methods: This is a retrospective analysis of eight patients and 75 additional cases from the literature. Data search included: patient's age, primary manifestation, risk factors, neuroimaging and outcome.

Results: Five female and three male patients gave consent to participate. Mean age was 12.5 years (10–15 years). Six patients presented with paraplegia; this was preceded by pain in four. Brown Sequard syndrome and quadriparesis were the two others' presenting condition. Sensation levels were thoracolumbar in seven cases. Bladder dysfunction only or bladder and bowel dysfunction were reported in eight and five patients respectively. Time to maximal symptom manifestation was <12 h in 7/8. Risk factors included surgery, minor trauma, recent infection, and thrombophilia. Mean follow-up was 3.3 years (0.25–6.3 years). Three patients remained wheelchair-dependent and three patients were ambulatory without aid. Bladder function recovered fully in five children. Most affected aspects of quality of life were physical and mental well-being and self-perception.

T2-weighted-MR images showed pencil-like hyperintensity (8/8) in sagittal and H-shaped or snake-eyes-like lesion (6/8) in axial views.

Analyses of all 83 patients were in congruence with the above results of the study group.

Conclusion: Spinal cord ischaemia in childhood presenting with pain, paraplegia, and bladder dysfunction has high morbidity concerning motor problems and quality of life. Acute arterial ischaemic event in children seems similar to adult events with respect to clinical presentation and, surprisingly, also in outcome.

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http://dx.doi.org/10.1016/j.ejpn.2012.09.002

1. Introduction

Spinal cord ischaemia (SCI) is a rare, but devastating event in childhood. There are no epidemiological data and only few literature reports on primary manifestation and outcome of SCI in both children and adults exist. Outcome data for children are very sparse consisting of case reports and children included in adult studies.[1–5] There is one small paediatric study from Blennow and Starck,[6] which showed recovery of bladder function in 6/7, full recovery of sensation in 3/7, and normal walking ability in 1/7 children. In another study by Sohal et al.,[7] a review of 20 cases in the paediatric literature (some of those patients are included in this study), two patients were dead at follow up (FU) and seven were walking with technical aids.

SCI is due to hypoperfusion or embolisation of the spinal artery, with the anterior spinal artery being most frequently affected.

Similar to cerebral arterial ischaemia in childhood, multiple risk factors are found in the literature. The most frequent provoking factors are minor trauma (with fibrocartilaginous embolus), parainfectious vasculopathies as well as aortic, cardiac, or spinal surgery.[8] Although, fibrocartilaginous embolism is considered to be a frequent risk factor, the diagnosis is proven only in a few lethal cases undergoing autopsy and histological examination.[9–11]

As mentioned above, the anterior spinal artery syndrome (ASAS) is the most common clinical presentation. These patients present with paraplegia/quadriplegia, bladder and/or bowel dysfunction and a sensory level with loss of pain and temperature sensation, but intact vibration sense and proprioception due to sparing of the posterior columns.

This is an observational retrospective study of a limited cohort collected from twelve Swiss neuropaediatric centres. The aim of this study is to describe the clinical manifestations, neuroimaging findings and outcome of SCI. Furthermore, we compare our observations with previous studies, and present a review of the relevant literature.

2. Patients and methods

2.1. Study patients

We retrospectively tried to relocate as many children as possible treated for SCI in Swiss children's hospitals.

Inclusion criteria were: (1) living in Switzerland at time of event, (2) aged one month to 16 years, (3) SCI or acute transverse myelopathy where SCI was most likely, but myelitis could not be ruled out completely.

SCI was defined in accordance to Masson[12]: acute myelopathies (development of symptoms in less than 27 h), no inflammatory signs in blood and cerebrospinal fluid (except discrete proteinorachia or pleiocytosis), presence of a risk factor for SCI and a neuroradiological lesion on MRI imaging in the territory of a spinal artery.

Exclusion criteria were acute transverse myelitis, demyelinating problems and high impact trauma (minor trauma provoking later ischaemia was accepted). Minor trauma includes all physical efforts triggering immediately progressive paraplegia or developing of neurological deficits within hours to days after an experience of back pain.

Relocation was achieved by contacting the twelve paediatric centres in Switzerland with a neuropaediatric division or neuropaediatric consult service. They were asked to retrieve all cases of SCI from their hospital data. Case retrieval was performed partly by data base search and partly by personal memory of the local neuropaediatricians. From the relocated patients data search was performed by studying the medical charts for age at manifestation, symptoms at manifestation, diagnostic investigations, risk factors and outcome.

2.2. Measures of outcome

Fourteen patients fulfilled inclusion criteria, but only nine patients could be relocated for the study. Finally eight gave informed consent.

All eight patients provided outcome data through questionnaires. One questionnaire was collecting data on the actual life situation, including information on residual symptoms, limitation of daily activities (data for Barthel-Index[13]), current therapies, subjective well-being, degree of recovery, and professional plans for the future. A second questionnaire was the standardised Kidscreen questionnaire about quality of life (QoL) for children from 8 to 18 years.[14,15] The Kidscreen questionnaire was completed by seven patients (two patients were older than 18 years). Patient 3 refused to fill out this questionnaire as she had just passed 20 years of age.

All patients were asked for a clinical FU including a detailed neurological examination and a short interview for completing information from medical data and questionnaires. Four patients (pat. 2,3,5,8) agreed. FU data for the others were taken from the last documented hospital visit.

The functions at FU were graded as follows: (1) normal function, (2) mild impairment: findings in neurological examination, not interfering with daily life activities, (3) moderate impairment: clinical impairment with difficulties in daily life, (4) severe impairment: loss of functions. The Barthel Index was calculated from the data given in the questionnaire (normal value = 100 points). Data on QoL was analysed for the ten domains in T-values (normal value $\geq$ 40).

MR imaging of spinal cord were reviewed by a neuroradiologist (MEK). Points of analysis included: (1) localisation and extension of spinal cord lesion, (2) signal changes on the sagittal and axial T2-weighted MR images, (3) pathological enhancement, (4) hyperintense signal on the diffusion-weighted image (DWI) in the affected segment and related ADC map. For the increased signal in the anterior horns of the grey matter the terms "snake-eyes" or "owl's eyes" sign have been used interchangeably in medical literature.[1,16–18]

2.3. Data collection from literature

A literature research for further cases was performed using the key words: 'spinal cord ischaemia', 'spinal cord infarction', 'myelopathy', 'anterior spinal artery syndrome' in online databases pubmed.com and embase.com. Additional reports

were identified in the reference lists of relevant articles. Inclusion und exclusion criteria for literature cases were the same as for the study patients, but the age criteria was expanded to 18 years, as this was usually the case in the literature. Relevant publications were dated from 1975 to 2010. We considered English, French, and German articles, where we had access to paper or electronic articles. Additional 75 patients were retrieved from the literature. Literature reports were searched through for: age at time of manifestation, clinical manifestation, MR finding, risk factors and FU data.

Statistical analyses were performed using SPSS version 18.0 and a p-value <0.05 was considered as significant. For analyses of categorical data Pearson chi-square tests were used and if expected frequencies were below five a Fisher's exact test was performed. The test of Kruskal Wallis and Mann–Whitney U-test were used for non-parametric data. Correlations were calculated by means of Spearman correlation coefficient r_s.

3. Results

3.1. Study patients

3.1.1. Presentation

Mean age at manifestation was 12.5 years (range ten to 15 7/12 years). All cases occurred between 2003 and 2009. Table 1 summarises findings at time of manifestation and FU. Patient 8 presented with diparesis in the arms, which evolved into quadriparesis after an episode of bradycardia and hypotension.

In Patients 6,7,8 minor trauma was reported (forced reclination, leapfrogging, practising somersault). Two of them suffering from pain immediately after trauma and developing hours to days later paraplegia. In one of them paraplegia developed right after trauma. An upper respiratory tract infection three weeks prior to the event was present in two children (pat. 2,5). Investigations for further risk factors revealed a homozygous MTHFR-C677T-Mutation in two patients (pat. 1,4) and a heterozygous MTHFR-A222V-Mutation in another patient (pat. 3). Multiple risk factors were present in patient 4 (homozygous MTHFR-mutation and spinal surgery).

3.1.2. Follow-up

Mean time to FU was 3.3 years, ranging from three months to 6.3 years, data is summarised in Table 1.

Four children (pat. 1,2,4,8) reported persistent pain problems due to spasticity and contractures ranking from 1 to 6 on a visual analogue scale from 0 to 10. Results of QoL are summarised in Table 2.

From four patients (pat. 1,4,5,7) rating their recovery as good, two (pat. 1,5) were still dependent on walking aids. Three children (pat. 2,3,8) reported some improvement, one of them was walking independently (pat. 8), the other two were still wheelchair-dependent. Patient 6, being wheelchair-dependent, did not consider himself as having made any improvement. None of the patients reported complete recovery nor complained about progressive symptoms. Physiotherapy, especially swimming, was considered to be the most helpful activity during rehabilitation (pat. 1,2,4,5,7,8).

At FU six children were attending school. Important to point out is that two patients (pat. 1,3) were neither in school nor involved in professional activities, for one of the them (pat. 3) there was a pre-existent psychiatric problem, which was more prominent.

3.1.3. Imaging

MR scans were on day one in all but one patient, patient 3 had a delayed initial MR on day two due to misdiagnosis of psychogenic paraplegia. Patient 1 was the only child with normal initial MR, but ischaemic anterior lesion was evident in an MRI on day 6. A "snake-eyes-like" configuration, i.e. band-like T2-hyperintensity between anterior horns (Fig. 1B), was prominent in five children (pat. 2,3,5,7,8) and an H-shaped T2-hyperintensity (Fig. 1C) in four cases (pat. 2,3,4,8) in axial imaging. In three (2,3,8) with band-like lesion in upper levels signal changed into H-shaped lesion in lower levels. The caudal parts of the lesions showed mostly irregular patterns, involving nearly the complete spinal cord section (see Fig. 1D). Typical snake-eyes sign was detected in three children in FU imaging as enhancement after gadolinium administration (Fig. 1A). All patients exhibited a vertical pencil-like T2-hyperintensity over multiple segments on sagittal imaging (see Fig. 2A; for radiological levels see Table 1). Patient 5 and 8 showed progression of the myelopathy within 2–7 days (see Fig. 2A–D). Patient 6 presented with a roundish T2-hyperintense lesion with focal areas of enhancement in the central part of the cord. Diffusion weighted imaging (DWI) was available in four patients (pat. 2,3,4,5). All of them showed hyperintense signal on DWI images (with maximal b-values).

3.2. Study patients and literature cases

Seventy-five patients[1–11,19–55] were retrieved from the literature. Together with our own study group of 8 patients, the data from these 83 combined cases was analysed.

3.2.1. Presentation

Mean age at presentation was 10.9 years, age and sex distribution are summarised in Fig. 3, showing a peak in adolescence and in preschool age. The sex ratio was M:F of 1.3:1.

Motor weakness was reported in all 83 children, detailed data was available in 70 patients: 45 (54%) were paraplegic/-paretic and 20 (24%) quadriplegic/-paretic. Three patients (4%) were reported to have hemiplegia due to Brown Sequard syndrome. Two patients presented with initial diplegia/-paresis of the arms which progressed within 12 h into quadriplegia/-paresis.

Five children (6%) had no sensory problems, for 13 (16%) no information was given. The sensation loss typical for ASAS (see introduction) was present in 31 (37%). Thirteen patients (16%) had loss of all qualities of sensation. There were 21 patients (25%) with other variable problems of sensation. Fig. 4 gives an overview of sensory levels. There is a mild predilection for lower thoracic and higher lumbar levels. Patients with systemic hypotension (surgery and/or low cardiac output) had a tendency to have ischaemic lesion between Th2–Th8 – sometimes referred as the watershed area of the spinal cord - but this finding was not statistically significant ($x^2(df = 3, n = 56) = 5.301$, $p = 0.151$).

Table 1 – Summery of clinical manifestation and FU of the study patients.

Patient	1	2	3	4	5	6	7	8
Age/sex	10 y/M	12 y/F	15 y/F	13 y/F	13 y/F	10 y/M	12 y/M	15 y/F
Clinical manifestation								
Pain	None	Lateral & abdominal	Leg	None	Thoracic back	None	Thoracic back	None
Motor impairment	Paraplegia	Paraplegia	Paraplegia	Plegia of right leg, paresis of left leg	Plegia of right leg, paresis of left leg	Paraplegia	Paresis of right leg and arm	Diparesis of arms (mainly left-sided)
Bladder and bowel dysfunction	Bladder & bowel dysfunction	Bladder dysfunction	Bladder & bowel dysfunction	Bladder & bowel dysfunction	Bladder & bowel dysfunction	Bladder dysfunction	Bladder & bowel dysfunction	Bladder dysfunction
Sensation impairment								
Level	L1	L1	L4	L2 right/Th9 left	Th4	L1	Th8 left	C3
Quality	All	All	All	Dysesthesias right/temp. & pain left	Temp., pain, dysesthesias	Temp.	Temp. & pain left	Dysesthesias
Time to max. symptoms	Unknown	>4 h, <12 h	<12 h	Postoperative	<4 h	>12 h, <24 h	>4 h, <12 h	1 h
Radiological lesion	Th10–CM	Th8–CM	Th11–CM	Th7–Th9	Th3–Th5	Th3–CM	C5–Th3	C3–Th2
Clinical syndrome	TM	TM	TM	Asymmetric ASAS	Asymmetric ASAS	ASAS	Brown Sequard	Not allocatable
Follow-up period	6 y 4 m	2 y 11 m	3 y 10 m	3 m	1 y 10 m	2 y 8 m	4 y 11 m	3 y 8 m
Outcome for…								
Motor impairment (ambulation)	Severe (walker)	Moderate (wheelchair)	Severe (wheelchair)	Moderate (ambulates unaided)	Moderate (orthosis)	Severe (wheelchair)	Normal	Minimal
Bladder incontinence	–	+	++	–	–	++	–	–
Bowel incontinence	–	–	–	+	–	++	–	–
Sensory problems	No information	Severe (pain, temp., touch)	Severe (temp., pain, touch)	Severe (temp.)	Moderate (touch left-sided)	No information	Severe (pain, temp.)	Severe (pain, temp.)
Barthel index	95	90	65	95	100	50	100	100

F: female; M: male; ASAS: Anterior spinal artery syndrome; TM: transverse myelopathy; y: years; m: month; h: hours; CM: conus medullaris; +: occasionally; ++: regularly; –: no; temp.: temperature.

Table 2 – T-values from Kidscreen quality of life for the study patients.

Patient	1	2	3	4	5	6	7	8
Physical well-being	**34**	61	–	**30**	**25**	**37**	44	**38**
Psychological well-being	44	**28**	–	**33**	40	**13**	54	**31**
Moods and emotion	45	47	–	**35**	42	**35**	53	42
Self-perception	50	48	–	**38**	**30**	**39**	71	**36**
Autonomy	45	46	–	42	**30**	51	58	**36**
Parent relation and home life	54	58	–	**34**	40	48	58	**32**
Peers and social support	48	52	–	**39**	45	53	52	45
School environment	64	53	–	44	51	**35**	60	**34**
Bullying	46	56	–	60	**30**	61	58	57
Financial resources	54	57	–	42	**22**	62	62	**18**

Bold values: below 40 (considered to be conspicuous).

Bladder dysfunction was reported in 50 patients (60%) in 29 of them together with bowel dysfunction. Two children (2%) had neither bowel nor bladder dysfunction. For 29 patients (35%) no information about autonomic disorders was available.

Pain was reported in 28 patients (34%). In the majority of patients pain was at the affected level, sometimes belt-like but also retrosternal, abdominal radiating in legs or groins. In half of the patients onset of pain was followed by a symptom-free interval (minutes up to twelve hours). Thirteen patients (16%) described dysesthesias often present as first symptoms together with pain. In 44 cases MR findings were reported. The radiological abnormalities always exceeded several levels. In 18 patients (41%) the lesion was described in the anterior or central cord. Two patients (5%) had a normal MR scan.

3.2.2. *Risk factor*

Table 3 gives a summary of all risk factors. For statistical analyses risk factors were allocated in four main groups as systemic hypotension (low cardiac output, all kind of surgery), inflammatory diseases (infections, parainfectious reactions and systemic diseases), minor trauma, and other risk factors.

Minor trauma seems to be a more frequent risk factor in adolescence and inflammatory disease and systemic hypotension in younger children (see Fig. 5). But there is no

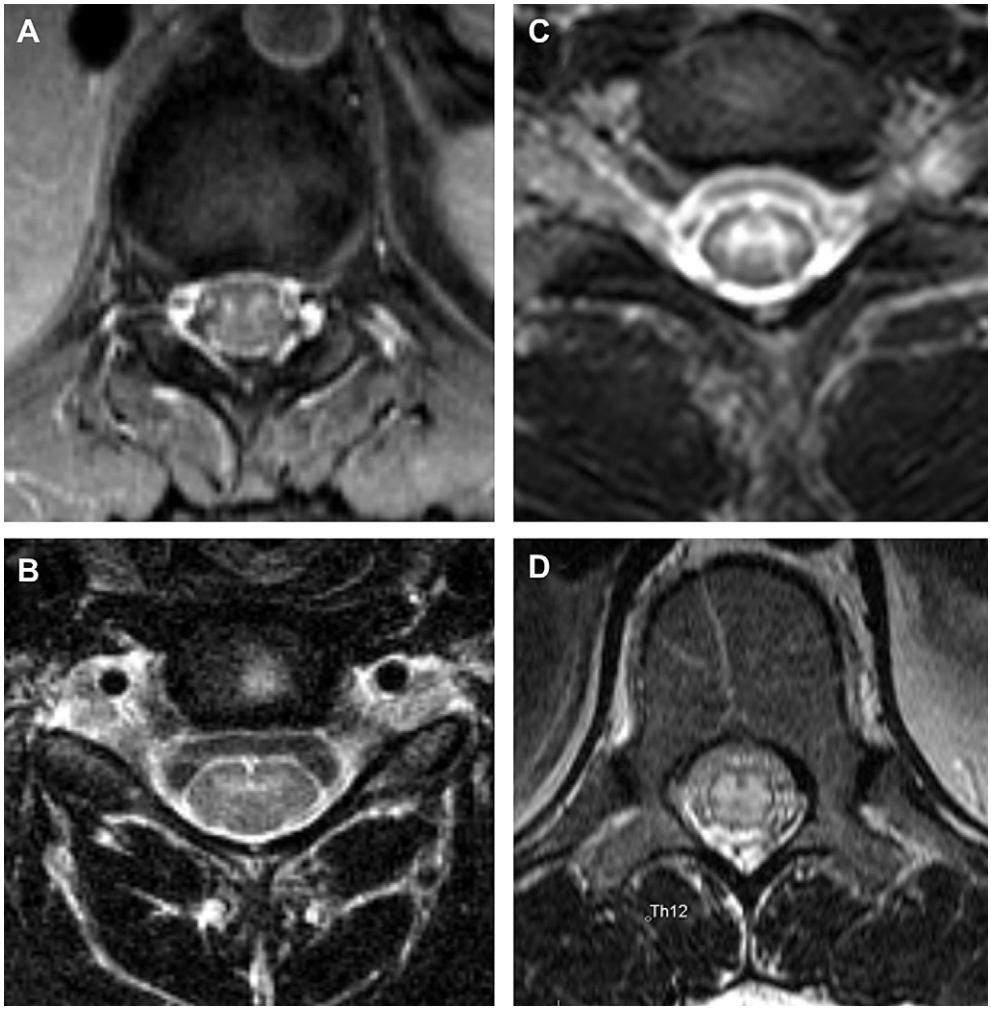

Fig. 1 – Various patterns of signal changes in spinal cord ischaemia: Snake-eyes configuration of enhancement after intravenous gadolinium in patient 3 (A). Snake-eyes-like (band-like) hyperintensity between the anterior horn in patient 8 (B). H-shaped T2 hyperintensity of the grey matter in patient 8 (C). Near complete involvement of the spinal cord section in patient 3 (D).

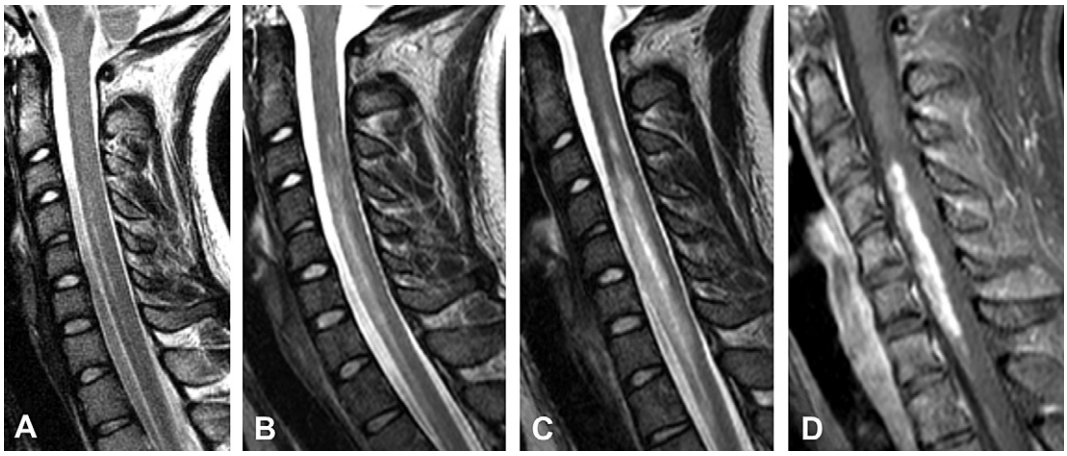

Fig. 2 – The development of the spinal cord ischaemia (patient 8): Sagittal T2-weighted images in day 1 (A) show a pencil-like vertical hyperintensity in the cervical spinal cord. On the following day (B) progressive swelling and hyperintensity is noted. One week later (C) further progression is seen accompanied by intramedullary enhancement after intravenous gadolinium (D).

statistical significance for this observation (Kruskal Wallis test; $x^2(df = 3, n = 79) = 3.742$, $p = 0.291$).

3.2.3. *Follow-up*

FU data were available for 74 children ranging from two week to 27 years after the event, in 24 cases time of FU was unknown.

Three of the 74 patients died (4%) six weeks to six months after the acute event. One died from respiratory arrest, one due to bronchial aspiration, and one of unknown cause. Four children (5%) recovered completely; nine (12%) patients had no improvement at all.

In all 74 patients information on motor weakness was given at FU. Five patients (6%) recovered completely, 54 (65%) had some recovery and 16 (18%) had no motor improvement at all. Of 44 patients with information on walking abilities, 14 (32%) were walking without aid, 23 (52%) with walking aids (such as orthosis, crutches, rollator, walker), seven (16%) were wheelchair-dependent.

FU on sensory function was given in 32 patients, 6 (19%) recovered completely, 16 (50%) partially and 10 (31%) had no improvement at all.

FU information on autonomic bladder and bowel symptoms was available for 43 children, 15 (40%) recovered completely, 7 (16%) partially with persistent problems such as necessity for intermittent catherization, 16 (37%) showed no improvement at all.

Complaints about chronic pain were only reported in six patients (8%).

In statistical analysis, there was no correlation between age at manifestation and functional outcome (motor: Spearman $r_s = -0.061$, $p = 0.607$, sensation: Spearman $r_s = 0.184$, $p = 0.314$ and autonomic symptoms: Spearman $r_s = 0.079$, $p = 0.633$) nor between the clinical level and outcome (motor: Spearman $r_s = -0.149$, $p = 0.313$ sensation: Spearman $r_s = -0.006$, $p = 0.976$ and autonomic symptoms: Spearman $r_s = -0.061$, $p = 0.607$).

There was no statistical significant difference of outcome of ambulation ($U = 210.5$, $z = -0.156$, $p = 0.876$) or death ($p = 0.14$) of cases published from 2000 to 2010 (study patients included) and from 1975 to 1990.

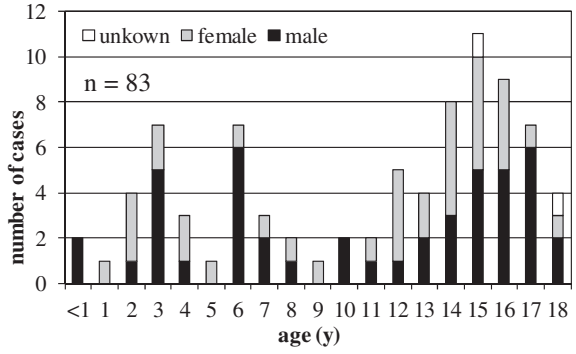

Fig. 3 – Age and sex distribution (study patients + literature cases).

4. Discussion

Together with our own thoroughly documented eight cases, we analysed data of 83 children with SCI.

The sex ratio shows a male predominance, which is slightly less prominent than in cerebral arterial ischaemic stroke.[56–58]

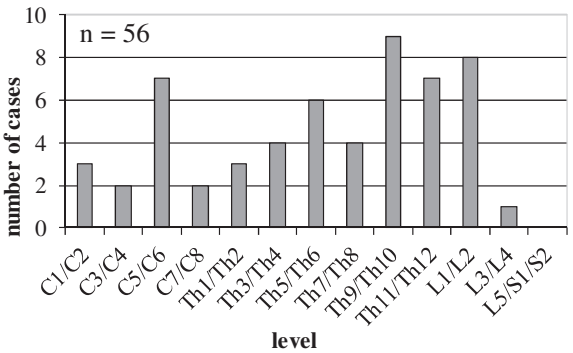

Fig. 4 – Sensory levels (study patients + literature cases).

Table 3 – Summary of risk factors. Multiple listing due to multiple risk factors in 25% of children.

Risk factor	*n*
Surgery	30
Minor trauma	22
Infection (meningitis)	15 (8)
Systemic hypotension	14
Hereditary thrombophilia	10
No obvious cause	4
Systemic diseases	3
Radiotherapy	2
Tumour	1
Cocaine misuse	1
Arteriovenous malformation	1

The prevalence of SCI is higher in adults than in children due to higher prevalence of atherosclerosis and aortic pathology which are major providers of SCI. Compared to reports from adults, children have similar symptoms and signs at manifestation. ASAS being by far the most frequent presentation (37% in our study compared to 37% in study of Novy et al.[59] and 67% in the study of Nedeltchev et al.[60]). This different prevalence of ASAS in the study of Nedeltchev et al. is most likely altered to more strict inclusion and exclusion criteria in this study. Based on anatomical studies the midthoracic level (Th4–Th8) has been thought to be the watershed area of the spinal cord with an increased vulnerability to ischaemic lesions.[2,4,62] Duggal and Lach[62] found ischaemic lesions in 95.5% of cases with hypotension or cardiac arrest in the lumbosacral levels. Duggal and Lach[62] stated that the midthoracic cord is the anatomic watershed area of the spinal cord, but that neurons in the lumbosacral levels seem to be more susceptible to ischaemia. This theory is supported by results of Cheshire et al.[4] who rejected as well this traditional hypothesis of the midthoracic cord being the watershed of the spinal cord. Cheshire et al.[4] also found the majority of ischaemic lesions localised to lumbosacral levels. In our study the ischaemic lesions are most frequent in the lower thoracic and lumbar cord (Fig. 4). Taking underlying risk factors for ischaemia into account, children with systemic hypotension were prone to midthoracic cord lesions, although no statistical significance could be shown ($p = 0.151$).

Masson et al.[63] described neurological problems in adults often being preceded by back or neck pain. Our study shows that one third of children initially presented with pain. In half of those presenting with pain, the pain was an isolated symptom which preceded further neurological symptoms up to twelve hours later.

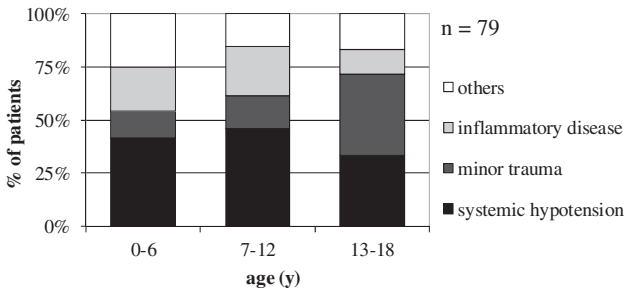

Fig. 5 – Risk factors for each age group (study patients + literature cases).

In a large review[8] (including neonatal cases) risk factors for SCI in children were: 26% minor trauma (fibrocartilaginous embolism included) and 19% parainfectious vasculopathies. This study did not mention any cases of spinal surgery and only a few cases of aortic surgery. However, in our series of the literature review surgery is the most prominent risk factor. It is not clear if Nance and Golomb[8] excluded SCI after spinal surgery. In more than a quarter (27%) of the children analysed by Nance and Golomb[8] more than one risk factor was present. In 39% of all children in this study at least one iatrogenic risk factor (such as surgery and radiotherapy) was present.

In contrast, to the current paediatric study, Nedeltchev et al.[60] showed arteriosclerosis (~33%) and aortic pathologies (33–43%) as most frequent risk factors in adults. Another study by Robertson et al.[64] found ischaemia perioperatively in 45% of adults, overwhelmingly from aortic surgery.

We found two peaks for SCI in childhood: preschool age (~2–6 years) and adolescents (~12–17 years; Fig. 3). Minor trauma seems to be more important in adolescence and infection, parainfectious reactions and systemic hypotension in the younger patients (Fig. 5). However these differences were not statistically significant ($p = 0.122$).

Comparison of young adults and children in outcome of cerebral stroke revealed a similar prognosis.[65] This also holds true for children with SCI: the percentage of adults regaining independent walking ranges from 11 to 41%,[60,61,64] in our study 32% reached independent walking ability and a further 52% could walk using aids. There is no evidence that age influences the outcome of SCI, as previously discussed in Nedeltchev et al.[60] and Iseli et al.[66] Several studies[5,17,60,61,64,66] found that functional outcome in adults depends on the degree of initial neurological deficits, in particular on motor deficits. Due to the paucity of information, we cannot make claims for the whole group of children. However, our small study group of eight children, those with paresis showed a better motor outcome than those with complete plegia (Table 1). Surprisingly, even in children there are 18%, who do not show any motor improvement.

As in the study of Masson et al.[17] we found no correlation between the level of the ischaemia and the outcome.

Prognosis for bladder and bowel dysfunctions seems to be more favourable. This was also observed in the paediatric case series of Blennow and Starck,[6] where six of seven children regained bladder control.

Unfortunately, there is no evidence that functional outcome in SCI is better now than 20 years ago ($p = 0.876$). The same applies for mortality ($p = 0.14$).

In the literature, pain is rarely reported at FU. Only in some studies[5,17,59,64] is pain mentioned, but approximately a third to a half patients in this studies suffered from pain at FU, which interfered with daily activities and gave rise to depressive symptoms.[5] In our small study group 50% and in the overall study 8% of patients suffered from pain at FU. It seems that pain is an underreported problem in the literature, which should be considered as a co-factor for reduced QoL. As pain is an important problem for long-term FU: careful evaluation and adequate treatment especially for spasticity and preservation of free range of movements for the joints is important.

MRI performed for the study patients revealed that none of the eight had traditional snake-eyes hyperintensity as described in the literature.[1,16–18] However, the snake-eyes sign can be present on the FU scans (more than seven days later) as enhancement after gadolinium administration. Again the snake-eyes-like and the H-shaped lesion occurred more frequently than the typical snake-eyes. These MRI signs are sometimes difficult to distinguish from each other and probably represent a spectrum of spinal cord involvement. Such MRI patterns, especially when combined with the clinical suspicion, should be considered as highly suggestive of SCI in children.

4.1. *Limitations of the study*

This is a retrospective case series, with a small number of cases. The probability in retrieving the majority of these children seems high, due to the aspects of rare diagnosis and serious impact. Nevertheless, some children in Switzerland with SCI certainly might have been missed. The differential diagnosis of myelitis and SCI is sometimes difficult. As children frequently have parainfectious neurological problems, some children with SCI might have been misdiagnosed as myelitis.

The heterogeneous study group mainly consisting of case reports from the literature posed difficulties in statistical analysis.

A weakness of the study is that definite diagnosis of minor trauma with consecutive fibrocartilaginous emboli can only be confirmed by post-mortem histological examination. Only in one patient out of three fibrocartilaginous emboli could be proven by autopsy.[44,54] Thus in most cases diagnosis remains a clinical suspicion. Furthermore in this retrospective study results concerning minor trauma might be influenced by recall-bias and the actual frequency of fibrocartilaginous emboli might be over- or under-estimated.

We realised that pain and sensory problems as significant problems in our study group, which was not reflected in the whole group. Thus, we assume that evaluation and report of sensory and pain problems are not adequate in many studies. This assumption is supported by the fact that even in our study group chart notes often did not mention the sensory and pain problems which the patients reported on careful FU.

5. Conclusion

SCI in childhood, presenting with pain, paraplegia and bladder dysfunction causes a high morbidity including motor, sensory and autonomic problems thus heavily affecting QoL. An H-shaped or snake-eyes-like T2-hyperintensity is highly suggestive for SCI. The outcome is not related to either age at manifestation nor level of functional lesion, but on severity of initial manifestation.

Acute arterial ischaemic spinal event in the paediatric population does not seem to differ from adults SCI; the children neither have less severe clinical presentation, nor better improvement. The difference from adult SCI is the multiplicity of risk factors in children.

Acknowledgements

We thank all patients, who supported this study with their participation. We thank Devjani Sengupta for carefully reviewing the English manuscript.

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