Epidemiology and clinical management of Conus-Cauda Syndrome and flaccid paraplegia in Friuli Venezia Giulia: Data of the Spinal Unit of Udine

Emiliana Bizzarini (1), Agostino Zampa (1), Cristina Malisan (1), Chiara Pinzini (1), Paolo Di Benedetto (1), Helmut Kern (2), Nicoletta Adami (3), Ugo Carraro (3)

(1) Spinal Unit, Department of Rehabilitation Medicine, ASS N° 4 "Medio Friuli", Udine, Italy; (2) Laboratory of Translational Myology of the University of Padova Interdepartmental Research Center of Myology & Interuniversitary Institute of Myology & Italian C.N.R. Institute of Neuroscience, c/o Department of Biomedical Sciences, University of Padova, Italy; (3) Ludwig Boltzmann Institute of Electrostimulation and Physical Rehabilitation, Department of Physical Medicine, Wilhelminenspital. Vienna, Austria.

Abstract

The Spinal Unit of the Department of Rehabilitation Medicine of Udine admits patients with Spinal Cord Injury (SCI) after discharge from Neurosurgery or Reanimation Divisions. Demographic data, etiology, duration of hospitalization, bladder managements, and mobility at admission and discharge were revisited from January 2000 to March 2008. We treated 49 complete Spinal Cord Injury patients (ASIA Impairment Scale A). The patients have an average age of 38.63 years (range 16 - 67 years). The ratio between Male and Female was 4 : 1, 40 men e 9 women. All the patients were classified complete respectively to the ASIA Impairment Scale, when no motor or sensory function was preserved in the sacral segment S4-S5. All patients were classified with the Ashworth scale from score 0 to 4.

All SCI patients have been treated with conventional rehabilitation strategies for enhancing lower limbs function. At the same time these patients had active exercise for upper limbs and training for daily mobility tasks (transferring, bed mobility and sitting) and functional electrical stimulation. Really we found some difficulties to electrically stimulate denervated muscles, but we believe this approach could give more chances to patients in their recovery. The hope is to realize walking assisted by FES or a combination of this with treadmill and biofeedback training in flaccid paraplegia even in a clinical ambit. This is a good opportunity to engage a long and wide road that leaded us to be partners in the Rise2-Italy Project. A result of this collaboration is the Case Report here presented of the first enrolled person.

.

Key Words: SCI, *Conus-Cauda Equina*, Functional Electrical Stimulation.

Basic Applied Myology 19 (4): 163-167, 2009

The spinal cord tapers and ends at the level between the first and second lumbar vertebrae in an average adult. The most distal part of the spinal cord is called the conus medullaris, its tapering end continues as the filum terminale. The upper border of the conus medullaris is often not well defined. Distal to this end of the spinal cord is a collection of nerve roots, which are horsetail-like in appearance and hence called the cauda equina (Latin for horse's tail). These nerve roots constitute the anatomic connection between the central nervous system (CNS) and the peripheral nervous system (PNS). They are arranged anatomically according to the spinal segments from which they originated and are within the cerebrospinal fluid (CSF) in the subarachnoid space with the dural sac ending at the level of second sacral vertebra. The conus medullaris constitutes part of the spinal cord (the distal part of the cord) and is in proximity to the nerve roots. Thus, injuries to this area often yield a combination of upper motor neuron (UMN) and lower motor neuron (LMN) symptoms and signs in the dermatomes and myotomes of the affected segments. The Conus-Cauda Equina Syndrome is characterized by flaccid paraplegia, sensory impairment, pain, as well as bladder and bowel areflexia, sexual dysfunctions. It is a rare disease, which generally affects adults.

We revisited 490 stories of SCI patients at their first admission to the Spinal Unit of Udine from January 2000 to May 2008. We selected the patients with spinal cord injury below D1, with a complete lesion or with a motor deficit at the lower limbs which required an intensive rehabilitation program. We selected 49 complete Spinal Cord Injury patients (ASIA Impairment Scale A) and 144 incomplete SCI patients (6 AIS B, 66 AIS C, 72 AIS D) after discharge from Neurosurgery and Intensive Care Unit.

All the patients were classified complete respectively to the ASIA Impairment Scale, when no motor or sensory function was preserved in the sacral segment S4-S5. At the beginning all inpatients with complete lesion had permanent bladder catheter and all of them passed through intermittent catheterism becoming outpatients.

About the patients classified as A at the AIS the average age was 38.63 years (range 16 - 67 years). The ratio between Male and Female was 4 : 1, according to International Literature (40 men e 9 women).

In our sample the etiology was traumatic in 46 patients, non-traumatic in 3 patients. Road accident was the main traumatic cause (30 pt).

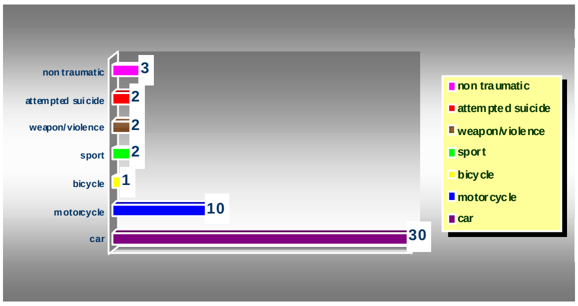


Table I: Etiology

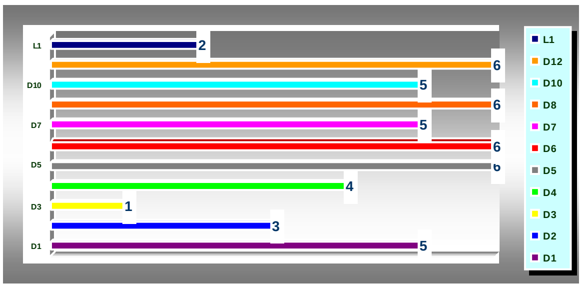
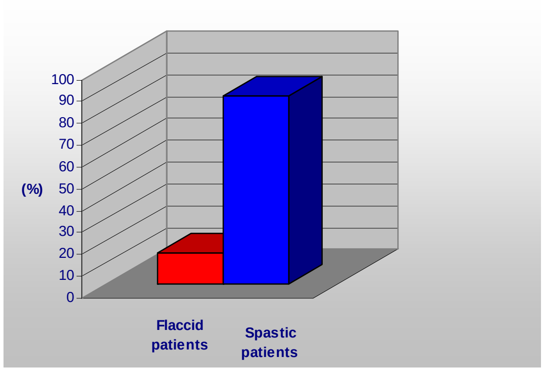


Table II: Level of Lesion

All patients were classified with the Ashworth scale (Ashworth, 1964) from score 0 to 4: 0 no increase in tone; 1 spastic catch; 2 more marked increase in tone but limb easily flexed; 3 considerable increase in tone, passive movement difficult; 4 limb rigid in flexion or extension ‘an ordinal level measure of resistance to passive movement’ [15].

About the patients classified with score 0, no increase in tone, at the Ashworth scale 1 had a lesion level at D2, 1 at D5, 3 at D12 and 2 at L1.



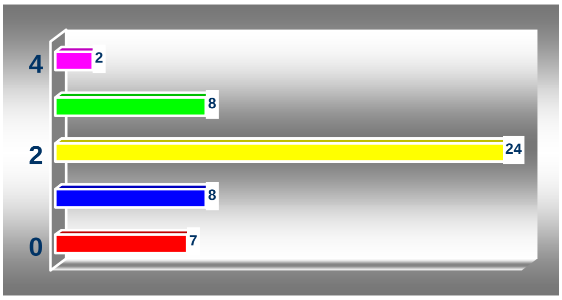


Table III: Ashworth score in SCI complete patients

Clinical management of flaccid paraplegia

All patients have been treated with conventional rehabilitation strategies for enhancing lower limb function after spinal cord injury focused on range of motion and stretching, upper limbs active exercise and functional training in daily mobility tasks (e.g. transferring, bed mobility, sitting) and electrical stimulation. The patients had active exercise for upper limbs and training for daily mobility tasks (transferring, bed mobility and sitting).

Current studies confirm positive results for the restoring of the locomotion in SCI as an intensive task-specific rehabilitative training, such as Body-weight Supported Treadmill Training (BWSTT), can promote supraspinal plasticity in the motor centers known to be involved in locomotion. Limited information, however, is provided describing the training progression, clinical decision making, and evaluation of progression steps. Our programs define a BWS initially set at 80% of BW. Initial training speed is chosen as the one most comfortable for the patient (0.1-0.2 m/s). Training intensity varies from 10 minutes to 45 minutes for session at a frequency of 5 days a week. The aims of the training are to decrease BWS and to increase speed in order to optimize the gait.

There are several available devices used for bracing the legs in order to support standing and walking function over ground, particularly for people with complete SCI. Among the most common long-leg braces studied in the literature are the purely mechanical Parawalker [16] or the Reciprocating Gait Orthosis (RGO) [5]. These devices must be used with a walking aid (e.g. crutches or walker) for functional ambulation.

The advantage of bracing is largely restricted to the general health and well being benefits related to practice of standing and the ability to ambulate short-distance in the home or indoor settings.

Energy expenditure of walking facilitated by bracing alone in spinal cord injury is extremely high and contributes to its low use.

Much research is focused on the development of effective therapies directed at enhancing locomotion.

Typically the majorities of these investigations focus on individuals with incomplete SCI and also predominately employ ambulation-related outcome measures. For the most part, these therapies include a form of body-weight supported treadmill training. Even if benefits can be obtained with conventional rehabilitation, locomotor training programs could be beneficial in improving lower limb muscle strength.

About our casuistry of complete SCI patients (N=49), locomotion training as therapeutic exercise was realized in 15 patients, beginning with Body-weight Supported Treadmill Training (BWSTT) and Knee Ankle Foot Orthosis (KAFO) personalized in soft-cast for the stabilization of the limbs. At discharge all patients used wheelchair for self transfer. We have experienced Patterned Electrical Stimulation assisted isometric exercise to prevent limb muscle atrophy. We know that PES-assisted isometric exercise reduces the degree of lower limb muscle atrophy in individuals with recent (~10 weeks post-injury) motor complete SCI, but not to the same extent as a comparable program of FES assisted exercise. Really a variety of electrical stimulation techniques are employed to enhance lower limb muscle structure in people with SCI. These typically involve delivering a series of electrical pulse trains to the muscle over time such that it simulates the “normal” exercise experience. Specific stimulation parameters (i.e., pulse width, train duration, between train intervals, method of application) and other exercise-related variables (i.e., frequency, duration, intensity, and program length) may each be varied to attain an optimal training stimulus.

Really we found some difficulties to electrically stimulate denervated muscles electrical but we believe this approach could give more chance to patients in their recovery. The hope is to realize walking assisted by FES also in combination with BWSTT and biofeedback training in flaccid paraplegia even in a clinical ambit. This is a good opportunity to engage a long and wide road that leaded us to be partners in the Rise2-Italy Project [1-4, 6-14].

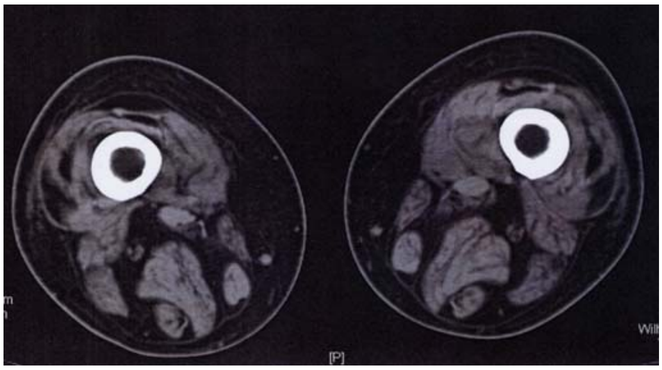


Fig. 1 *CT scan demonstrate that the healty macroscopic apperance of the thigh of Rise2-Friuli-01LS is misleading, being due to a typical pseudo-hypertrohy related to lipodystrophy of the denervated thigh muscles. The thick layer of subcutaneous adipose tissue, toghether with those separating denervated quadriceps and hamstring muscles is at least in part responsible of the difficulties encountered in leg muscle stimulation using commercial electrical stimulators for innervated muscles. Adipose tissue also infiltrate the atrophic muscles, and thus the compact aspects of normal thigh muscles is lost.*

Case Report

We report a 41-year-old male patient who sustained a severe traumatic spinal cord injury in an accident at work (April 11, 2008). On clinical examination he presented with flaccid paraplegia (motor level D12). Initial magnetic resonance imaging showed a fracture-dislocation D12-L1. After discharge from Neurosurgery at 15 days from trauma he was admitted in our Spinal Unit. At the clinical examination was evaluated as A at the ASIA Impairment Scale (Motor level D12, anesthesia below D12). From an electrophysiological point of view somato-sensory evoked potentials and motor evoked potentials were silent. He had begun the rehabilitation program with the purpose to reach autonomy in wheelchair locomotion and ADL. The physical rehabilitation was directed to the strengthening of upper limbs, trunk control and to preventing lower limb muscle atrophy with patterned Electrical Stimulation (quadriceps and tibialis anterior). Specific stimulation parameters (i.e., pulse width, train duration, between train intervals, method of application) were studied, but we cannot obtain an optimal training stimulus. The training was completed by passive exercise at the cycloergometer and assisted body-weight support treadmill training supported by FES. Gait over ground was not possible. At demission we verified autonomy in wheelchair locomotion and ADL, a good trunk control and an increment of aerobic performance (VO_{2max}). Meantime

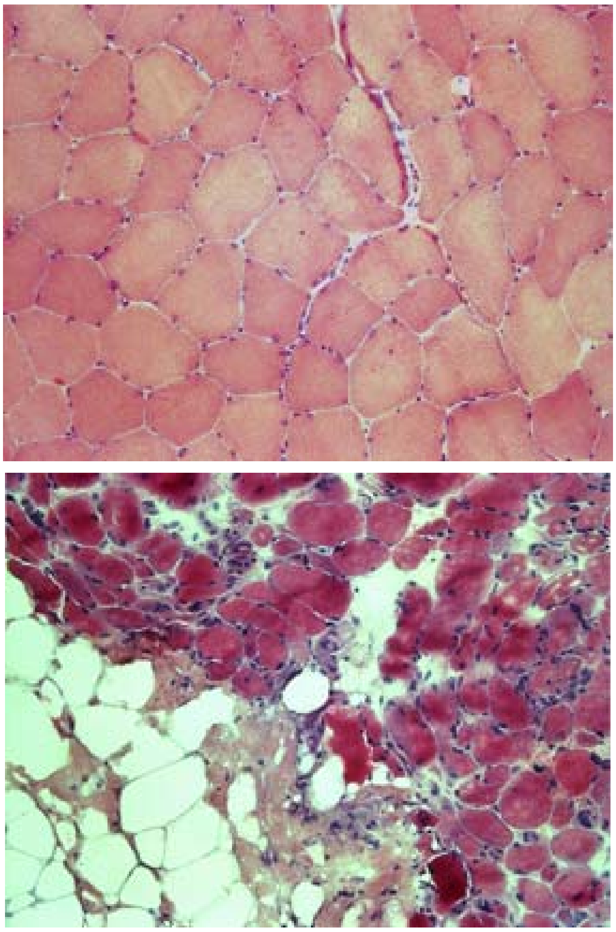


Fig 2 *Human quadriceps muscle. Upper panel: normal adult. Lower panel: Muscle biopsy of Rise2-Friuli_01LS at enrolment. Nine months after SCI the muscle fibers are atrophic and the interstitial tissue is increased. In the left lower corner a group of adipocytes of the infiltrating fat tissue are present. Hematoxilin-Eosin. 20x,*

some reinnervation of leg muscles occurred demonstrated by minimal spontaneous activity of right foot muscles, but voluntary or electrical stimulation-induced muscle contractions did not appeared.

On the other hand, ES-induced dorsiflection of the ankle were achieved by transcutaneous electrical stimulation of the tibialis anterior using the “Denervated Muscle Stimulation” program of the STIWELL Med4 device of MED-EL (Innsbruck, Austria), that is delivering triangular or bidirectional impulses of 70 maps intensity and 150 msec duration. After this demonstration of maintained muscle contractility, the patient accepted the burden to travel to Vienna (January 21-23, 2009), to be analyzed and enrolled in the 08Rise2-Italy Program after signing the Consent and have performed a Computer Tomography analysis of his leg muscle that allowed to measure the thick subcutaneous fat layer and to evaluate the extent of muscle degeneration that underwent during the 9 months since Spinal Cord Injury. CT scan of left and right leg of Rise2-Friuli-01LS demonstrates that the healthy macroscopic appearance was misleading, being due to overweight and to pseudo-hypertrophic lipodystrophy of long-lasting denervated thigh muscles (Figure 1). The thick layer of subcutaneous adipose tissue, together with those separating denervated quadriceps and hamstring muscles is in part responsible of the difficulties encountered in leg muscle stimulation using commercial electrical stimulators for innervated muscles. Adipose tissue also infiltrates the atrophic muscles, and thus the compact aspect of normal thigh muscles is lost.

Figure 2 shows the histological aspects of normal adult quadriceps muscle (upper panel) and the atrophic muscle of Rise2-Friuli-01LS (lower panel). After 9 months of permanent denervation muscle fibers are atrophic and the interstitial tissue is increased, but the fast and slow fiber types are still distinct by ATPase histochemistry (manuscript in preparation). In the left lower corner groups of adipocytes of the infiltrating fat tissue are present. Morphometry of muscle biopsies confirms that the muscle fibers have the characteristics of this stage of permanent denervation, while Functional Echomyography of Tibialis Anterior that the muscles are poorly perfused at rest (Manuscript in preparation). From March 2009 LS is performing Therapeutic Electrical Stimulation (TES) for denervated leg muscles, using custom-designed electrodes and stimulators developed in Vienna, Austria for the EU Project RISE (Contract n. QLG5-CT-2001-02191) [3, 10]. An early result is the impressive reduction of the leg edema.

This first experience strongly suggests that more subjects affected with lower motor neuron denervation of skeletal muscles than those enrolled in the EU Project RISE will benefit of the Vienna Strategy for Therapeutic Electrical Stimulation, in particular those difficult-to-stimulate due to thick subcutaneous adipose layer that hamper excitation of denervated or poorly innervated muscles.

Acknowledgements

The study was supported by funds of Interdepartmental Research Center of Myology of the University of Padova and Research Funds from the Ludwig Boltzmann Institute for Electrostimulation and Physical Rehabilitation at the Institute of Physical Medicine and Rehabilitation (Wilhelminenspital, Vienna, Austria).

Address Correspondence to:

Emiliana Bizzarini, Unità Spinale, Dipartimento di Medicina Riabilitativa, Azienda per i Servizi Sanitari N° 4 “Medio Friuli”, Udine, Italia
E-mail: biomec.imfr@ass4.sanita.fvg.it

References

[1] Bijak M, Mayr W, Rakos M, Hofer C, Lanmüller H, Rafolt D, Reichel M, Sauermann S, Schmutterer C, Unger E, Russold M, Kern H. The Vienna functional electrical stimulation system for restoration of walking functions in spastic paraplegia. Artif Organs 2002; 26(3): 224-7.

[2] Bijak M, Rakos M, Hofer C, Mayr W, Strohhofer M, Raschka D, Kern H. Stimulation parameter optimization for FES supported standing up and walking in SCI patients. Artif Organs 2005; 29(3): 220-3.

[3] Boncompagni S, Kern H, Rossini K, Mayr W, Carraro U, Protasi F. Structural differentiation of skeletal muscle fibres in absence of innervation in humans. Proc Nat Acad Sci USA 2007; 104: 19339-44.

[4] Carraro U, Rossini K, Mayr W, Kern H. Muscle fiber regeneration in human permanent lower motoneuron denervation: relevance to safety and effectiveness of FES-training, which induces muscles recovery in SCI subjects. Artif Organs 2005; 29(3):187-91.

[5] Douglas R, Larson PL, D'Ambrosia R, McCall RE. The LSU reciprocating gait orthosis. Orthopaedics 1983; 6: 834-9.

[6] Gallasch E, Rafolt D, Kinz G, Fend M, Kern H, Mayr W. Evaluation of FES-induced knee joint moments in paraplegics with denervated muscles. Artif Organs 2005; 29(3): 207-11.

[7] Graupe D, Cerrel-Bazo H, Kern H, Carraro U. Walking performance, medical outcomes and patient training in FES of denervated muscles for ambulation by thoracic-level complete paraplegics. Neurol Res 2008; 30(2): 123-30.

[8] Hofer C, Mayr W, Stöhr H, Unger E, Kern H. A stimulator activation of denervated muscles. Artif Organs 2002; 26(3): 276-9.

[9] Kern H, Hofer C, Mödlin M, Forstner C, Mayr W, Richter W. Functional electrical stimulation (FES) of long-term denervated muscles in humans: Clinical observations and laboratory findings. Basic Appl Myol 2002; 12: 291-7.

[10] Kern H, Boncompagni S, Rossini K, et al. Long-term denervation in humans causes degeneration of both contractile and excitation-contraction coupling apparatus that can be reversed by functional electrical stimulation (FES). A role for myofiber regeneration? J Neuropath Exp Neurol 2004;63:919–931.

[11] Kern H, Hofer C, Moedlin M, Forstner C, Raschka-Hoegler D, Mayr W, Stoehr H. Denervated muscles in humans - limitations and problems of currently used functional electrical stimulation training protocols. Artif Organs 2002; 26: 216-8.

[12] Kern H, Rossini K, Carraro U, Mayr W, Vogelauer M, Hoellwarth U, Hofer C. Muscle biopsies show that FES of denervated muscles reverses human muscle degeneration from permanent spinal motoneuron lesion. J Rehabil Res Dev 2005; 42 (3S1): 43-53

[13] Kern H, Hofer C, Modlin M, Mayr W, Vindigni V, Zampieri S, Boncompagni S, Protasi F, Carraro U. Stable muscle atrophy in long term paraplegics with complete upper motor neuron lesion from 3- to 20-year SCI. Spinal Cord 2008; 46(4): 293-304.

[14] Mayr W, Hofer C, Bijak M, et al. Functional Electrical Stimulation (FES) of denervated muscles: existing and prospective technological solutions. Basic Appl Myol 2002; 12: 287-90.

[15] Pandyan AD, Johnson GR, Price CI, Curless RH, Barnes MP, Rodgers H. A review of the properties and limitations of the Ashworth and modified Ashworth Scales as measures of spasticity. Clin Rehabil. 1999; 13(5) :373-83.

[16] Rose GK. The principles and practice of hip guidance articulations. Prosthet Orthot Int 1979; 3: 37-43.