# Malformazione di Chiari e Siringomielia

***Ruolo degli esami neurofisiologici***


**Marcello Romano**

**UOC di Neurologia e Stroke Unit**

**Servizio di Neurofisiopatologia**

**CRR distonie e tossina botulinica**

**AOOR Villa Sofia Cervello Palermo**


**Malformazione di Chiari e siringomielia: ruolo degli esami neurofisiologici**

il problema non può essere ridotto a una lettura puramente morfologica della risonanza magnetica

La **siringomielia** non è soltanto una cavità fluida all'interno del midollo, ma **l'espressione di una mielopatia dinamica**, nella quale la topografia della lesione determina il tipo di deficit clinico e, di conseguenza, il profilo neurofisiologico osservabile

la siringa non è solo un'immagine: è una lesione che, a seconda di come si dispone nel midollo, seleziona sistemi anatomico-funzionali diversi e genera per questo fenotipi neurofisiologici differenti.

Journal of Clinical Medicine

MDPI

*Review*

# A Critical Update of the Classification of Chiari and Chiari-like Malformations

**Juan Sahuquillo [1,2,3,*], Dulce Moncho [2,4], Alex Ferré [2,5], Diego López-Bermeo [1,2], Aasma Sahuquillo-Muxi [2] and Maria A. Poca [1,2,3]**

1 Department of Neurosurgery, Vall d'Hebron University Hospital, Passeig Vall d'Hebron 119-129, 08035 Barcelona, Spain; diegofernando.lopez@vallhebron.cat (D.L.-B.); pocama@neurotrauma.net (M.A.P.)
2 Neurotraumatology and Neurosurgery Research Unit, Vall d'Hebron Institut de Recerca (VHIR), Vall d'Hebron Barcelona Hospital Campus, Passeig Vall d'Hebron 119-129, 08035 Barcelona, Spain; dulce.moncho@gmail.com (D.M.); alejandro.ferre@vallebron.cat (A.F.); aasmasm@outlook.com (A.S.-M.)
3 Department of Surgery, Universitat Autònoma de Barcelona, Bellaterra, 08193 Barcelona, Spain
4 Clinical Neurophysiology Department, Vall d'Hebron Hospital Universitari, Vall d'Hebron Barcelona Hospital Campus, Passeig Vall d'Hebron 119-129, 08035 Barcelona, Spain
5 Sleep Unit, Pneumology Department, Vall d'Hebron Hospital Universitari, Vall d'Hebron Barcelona Hospital Campus, Passeig Vall d'Hebron 119-129, 08035 Barcelona, Spain
* Correspondence: sahuquillo@neurotrauma.net

**Table 1.** A summary of the different types of Chiari malformations (CMs) described in the indexed peer-reviewed literature under the eponym CMs. Due to the exceptionally rare anomalies and the absence of precise embryological investigations, CM3, CM3.5, CM4, and CM5 may be malformations unrelated to the more common CM0, CM1, CM1.5, and CM2. The versions that are kept in the ICC-CM classification are noted in bold and yellow background.

| Type | Abbreviation | Author \| Year | Main Characteristic |
|---|---|---|---|
| **Chiari 0** | **CM0** | Iskandar et al., 1998 [1] | Posterior fossa crowding. No TH |
| Chiari 0.5 | CM0.5 | Morgenstern et al., 2020 [2] | Ventrolateral tonsillar wrapping |
| **Chiari 1** | **CM1** | Chiari, 1891 [3] | Tonsillar herniation (>3–5 mm) |
| **Chiari 1.5** | **CM1.5** | Tubbs et al., 2004 [4] | Cerebellar and brainstem herniation |
| **Chiari 2** | **CM2** | Chiari, 1891 [3] | Spina bifida \| Cerebellar-brainstem herniation |
| Chiari 3 | CM3 | Chiari, 1891 [3] | Cerebellar-cervical encephalocele |
| Chiari 3.5 | CM3.5 | Fisahn et al., 2016 [5] | Cerebellar-cervical encephalocele connected to foregut |
| Chiari 4 | CM4 | Chiari, 1895 [6] | Cerebellar hypoplasia or aplasia. No TH |
| Chiari 5 | CM5 | Tubbs et al., 2012 [7] | Cerebellar aplasia and occipital lobe herniation |

**Table 2.** Etiological classification of tonsillar descent and Chiari-like disorders modified from Raybaud and Jallo [6].

| Category | Etiology | Pathologies |
|---|---|---|
| **Tonsillar herniation** | Increased volume \| High ICP | • High ICP<br>• Space-occupying lesions in the PF<br>• Diffuse brain swelling \| Brain edema.<br>• Intracranial hematomas<br>• Hydrocephalus, arachnoidal cysts |
| **Tonsillar herniation** | Cranio-spinal gradients | • Lumbar drainage<br>• Lumbo-peritoneal shunts<br>• Chronic spinal CSF leaks |
| **Primary Chiari malformations** | Mesodermal insuficiency | • CM0<br>• CM1<br>• CM1.5<br>• CM2<br>• Complex Chiari malformations |
| **Complex CVJ malformations** | Embriological mesodermal abnormalities | • Basilar impression, platybasia<br>• Clivus hypoplasia<br>• Retrocurved odontoid<br>• Occipitalization of the atlas<br>• Klippel-Feil syndrome |
| **Secondary Chiari-like malformations** | Secondary reduced volume of the skull or neural overgrowth | • Syndromic craniosynostosis<br>• Non-syndromic craniosynostosis<br>• Extrathecal shunting.<br>• Overgrowth syndromes (Costelo, Sotos, MCAP, etc.)<br>• Non-syndromic macrocerebellum |

Journal of
*Clinical Medicine*

MDPI

*Review*

# A Critical Update of the Classification of Chiari and Chiari-like Malformations

**Juan Sahuquillo [1,2,3,*], Dulce Moncho [2,4], Alex Ferré [2,5], Diego López-Bermeo [1,2], Aasma Sahuquillo-Muxi [2] and Maria A. Poca [1,2,3]**

1 Department of Neurosurgery, Vall d'Hebron University Hospital, Passeig Vall d'Hebron 119-129, 08035 Barcelona, Spain; diegofernando.lopez@vallhebron.cat (D.L.-B.); pocama@neurotrauma.net (M.A.P.)
2 Neurotraumatology and Neurosurgery Research Unit, Vall d'Hebron Institut de Recerca (VHIR), Vall d'Hebron Barcelona Hospital Campus, Passeig Vall d'Hebron 119-129, 08035 Barcelona, Spain; dulce.moncho@gmail.com (D.M.); alejandro.ferre@vallebron.cat (A.F.); aasmasm@outlook.com (A.S.-M.)
3 Department of Surgery, Universitat Autònoma de Barcelona, Bellaterra, 08193 Barcelona, Spain
4 Clinical Neurophysiology Department, Vall d'Hebron Hospital Universitari, Vall d'Hebron Barcelona Hospital Campus, Passeig Vall d'Hebron 119-129, 08035 Barcelona, Spain
5 Sleep Unit, Pneumology Department, Vall d'Hebron Hospital Universitari, Vall d'Hebron Barcelona Hospital Campus, Passeig Vall d'Hebron 119-129, 08035 Barcelona, Spain
* Correspondence: sahuquillo@neurotrauma.net

la siringomielia va interpretata all'interno di un sistema: quello della fossa posteriore, della giunzione cranio-cervicale e dei gradienti pressori liquorali. In altri termini, la siringa non è un reperto da leggere isolatamente, ma una conseguenza funzionale di un equilibrio alterato tra contenitore osseo, contenuto neurale e dinamica del liquor

# Ragionamento clinico preoperatorio

Nei pazienti con **Chiari isolata**, la decisione non può dipendere soltanto dalla presenza dell'ectopia tonsillare, ma deve integrare **sintomi, segni neurologici, anatomia della fossa posteriore ed eventuale evidenza di sofferenza funzionale**

Quando compare una **siringomielia**, però, il peso del ragionamento cambia: non si sta più valutando solo una malformazione della giunzione cranio-cervicale, ma **una vera lesione intramidollare potenzialmente evolutiva**. In questo contesto **assumono rilievo l'estensione longitudinale della cavità, la sua eventuale eccentricità, la presenza di siringobulbia, il decorso clinico e il tipo di deficit neurologico**

La neurofisiologia diventa allora utile non tanto per "confermare" la RM, quanto per chiarire quale sistema sia già funzionalmente compromesso e quanto la lesione sia biologicamente attiva

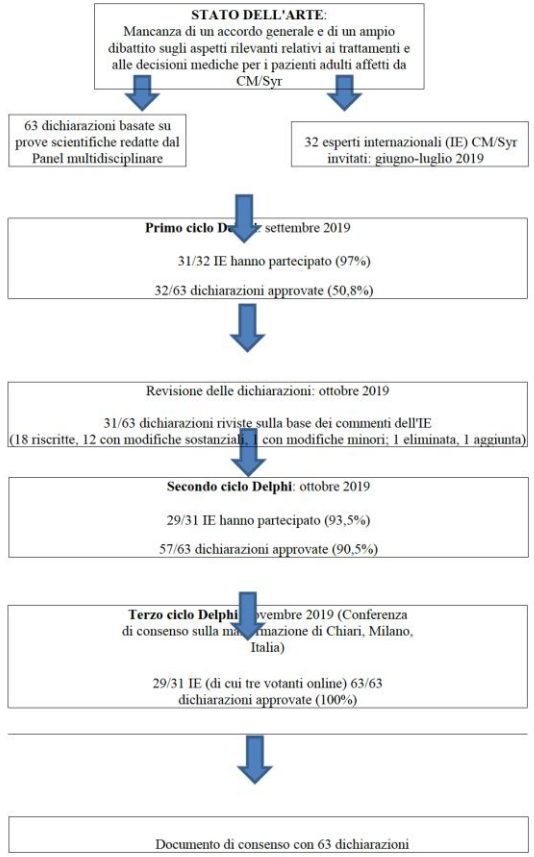
Neurological Sciences (2022) 43:1327–1342
https://doi.org/10.1007/s10072-021-05347-3

ARTICOLO ORIGINALE

# Diagnosi e trattamento della malformazione di Chiari e della siringomielia negli adulti: documento di consenso internazionale

Palma Ciaramitaro[1,2] • Luca Massimi[3] • Alessandro Bertuccio[4] • Alessandra Solari[5] • Mariangela Farinotti[5] • Paola Peretta [6] • Veronica Saletti [7] • Luisa Chiapparini [8] • Andrea Barbanera [4] • Diego Garbossa [1] • Paolo Bolognese [9] • Andrew Brodbelt [10] • Carlo Celada [11] • Dario Cocito [12] • Marcella Curone [13] • Grazia Devigili [14] • Alessandra Erbetta [15] • Marilena Ferraris [16] • Marika Furlanetto [17] • Mado Gilanton [18] • George Jallo [19] • Marieta Karadjova [20] • Jorg Klekamp [21] • Fulvio Massaro[22] • Sylvia Morar[23] • Fabrice Parker[23] • Paolo Perrini[24] • Maria Antonia Poca [25] • Juan Sahuquillo[25] • Marcus Stoodley[26] • Giuseppe Talamonti[27] • Fabio Triulzi [28] • Maria Consuelo Valentini [16] • Massimiliano Visocchi [29] • Laura Valentini [17] • A nome della Giuria Internazionale di Esperti della Conferenza di Consenso sulla Siringomielia di Chiari, Milano, 11-13 novembre 2019

Fig. 1 Bozza del processo di consenso




dati del registro Piemonte–Valle d'Aosta

la siringomielia presentava una prevalenza di 4,84/100.000 e un'incidenza di 0,82/100.000, ma soprattutto le forme sintomatiche risultavano frequenti e associate a disturbi sensitivi, deficit motori, dolore neuropatico, coinvolgimento dei nervi cranici e disfunzione vescicale

la siringomielia non è un reperto raro e "silenzioso" nel senso clinico del termine, ma una condizione che può produrre un carico neurologico significativo. Il semplice diametro della cavità non è di per sé sufficiente a definire la gravità del caso: ciò che conta è sempre il rapporto tra topografia della lesione e sistemi coinvolti.

All'interno di questo inquadramento, la diagnosi differenziale conserva un ruolo non secondario.

## Annex A. D.G.R. 95-13748 - 29/03/2010
## Diagnostic protocol

REGIONE PIEMONTE

- **Mandatory:**

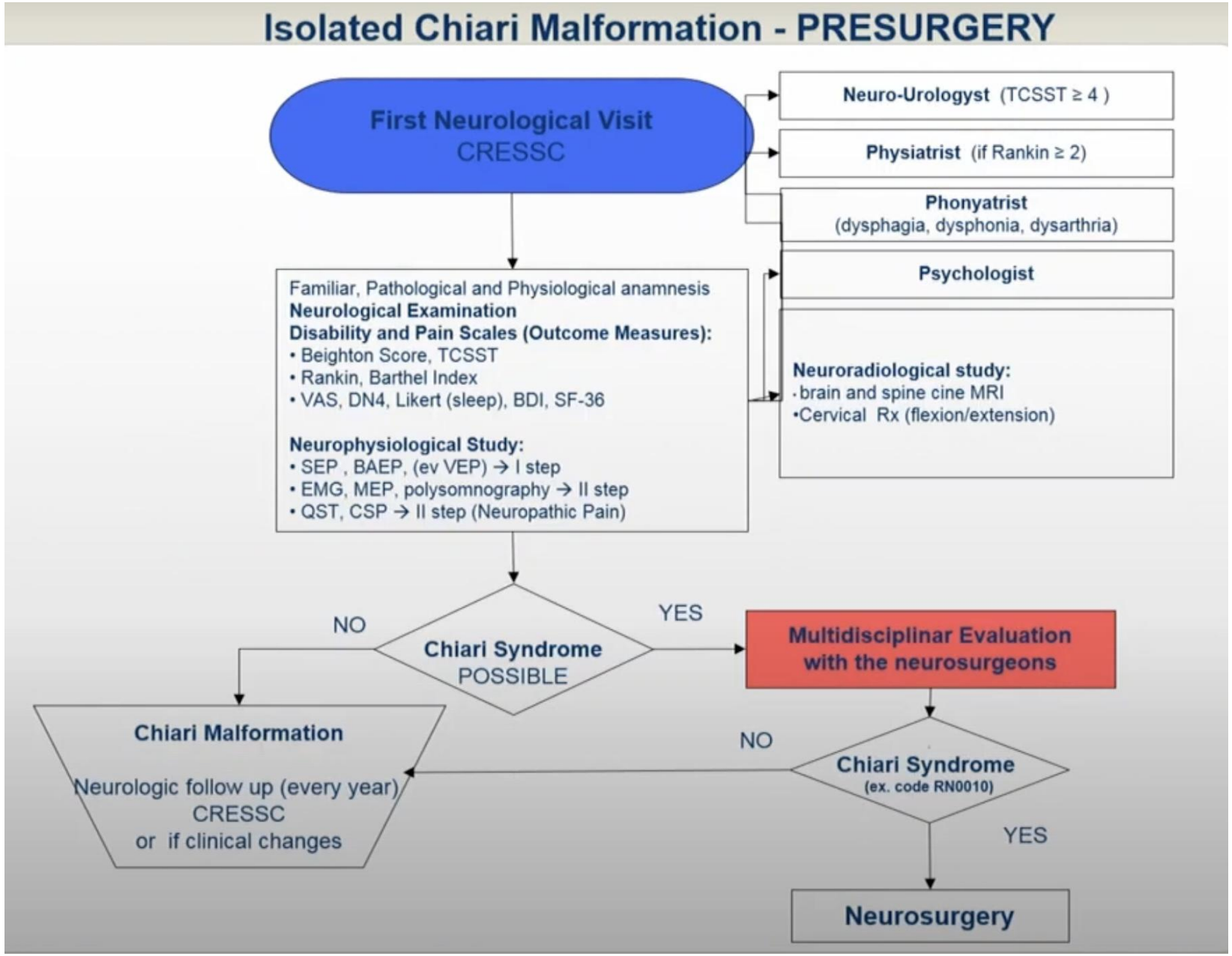
1. Neurological examination
2. Cine MRI brain and whole spinal cord

- **Additional tests**

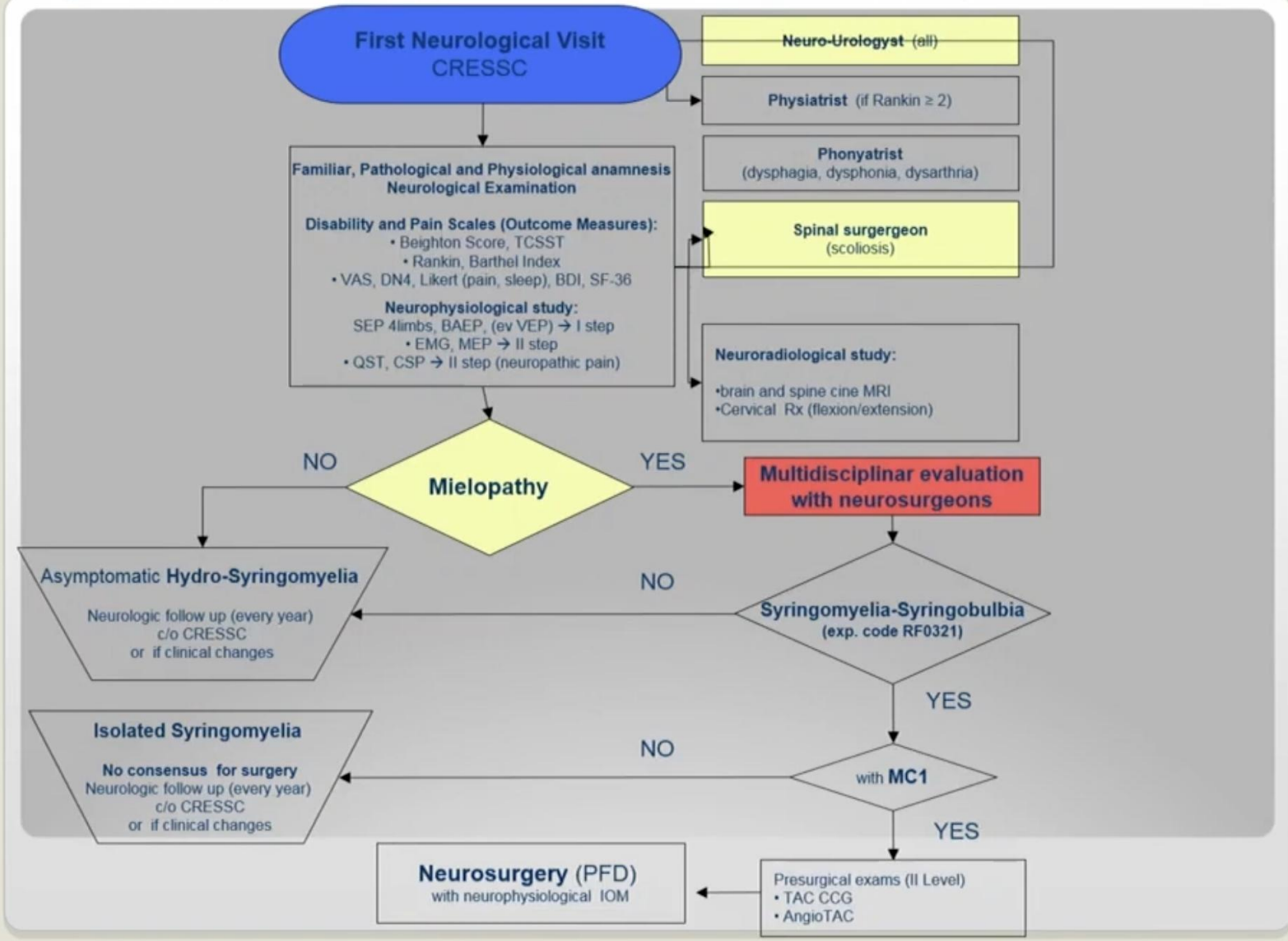
(clinical phenotype and severity definition):

1. EMG, SEPs, BAEPs, MEPs;
2. neurophysiological study of neuropathic pain (CSP, LEPs);
3. EEG, polysomnography (if suspected epilepsy, sleep apnea)

# Isolated Chiari Malformation - PRESURGERY



# Syringomyelia-Syringobulbia (with or withour MC1)-PRESURGERY



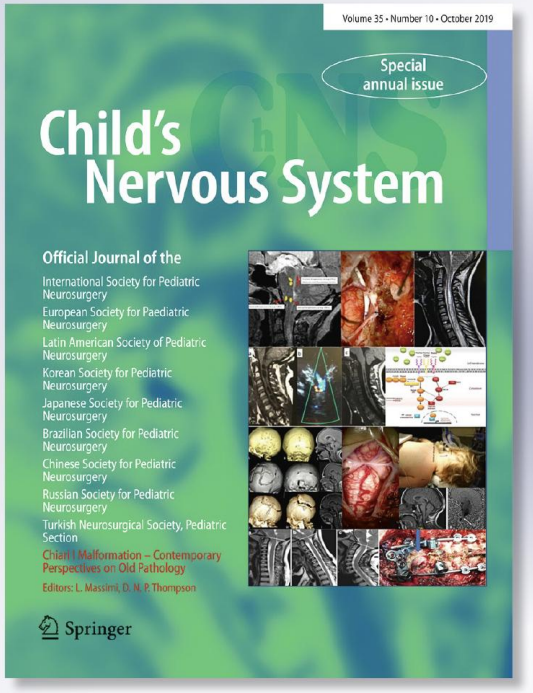
*Clinical diagnosis—part I: what is really caused by Chiari I*

**Palma Ciaramitaro, Marilena Ferraris, Fulvio Massaro & Diego Garbossa**

**Child's Nervous System**

ISSN 0256-7040
Volume 35
Number 10

Childs Nerv Syst (2019) 35:1673-1679
DOI 10.1007/s00381-019-04206-z




Springer

Child's Nervous System (2019) 35:1673–1679
https://doi.org/10.1007/s00381-019-04206-z

SPECIAL ANNUAL ISSUE

Check for updates

# Clinical diagnosis—part I: what is really caused by Chiari I

Palma Ciaramitaro[1] · Marilena Ferraris[2] · Fulvio Massaro[3] · Diego Garbossa[3]

Received: 18 April 2019 / Accepted: 9 May 2019 / Published online: 3 June 2019
© Springer-Verlag GmbH Germany, part of Springer Nature 2019

## Abstract

**Purpose** Chiari malformation is a group of congenital malformations involving the brainstem, cerebellum, and upper spinal cord, frequently identified in both young adults and in children. Chiari I malformation (CM1), classically defined as a caudal displacement of the cerebellar tonsils through the foramen magnum into the spinal cord, is the most common clinical type. A syringomyelia can be associated at the time of the diagnosis or appear secondarily and manifest with medullary symptoms. The aim of this paper is to update the knowledge on clinical manifestations specifically related to Chiari I malformation with or without syringomyelia in the pediatric population.

**Methods** Current literature with focus on relevant clinical pediatric issues is reviewed and discussed, comparing with those related to adults; we include the results of a 10-year single-center experience on 600 CM1 patients.

**Results and conclusions** Herniation of the cerebellar tonsils may lead to significant clinical symptoms, including neck and cervical pain, short-lasting occipital "cough" headache, dizziness, and gait impairment; in children younger than 3 years, oropharyngeal symptoms are prevalent (sleep apnea, feeding problems) whereas in those older than 3 years, a higher incidence of cough headache and scoliosis is reported. CM1 clinical features, both in children and in adults, have in common the presence of anatomical deformities of the brainstem and cerebellum. Clinical myelopathy (sensory/autonomic disorders, motor weakness) can result from direct compression of the cervical spinal cord by the herniated cerebellar tonsils or can be due to the presence of a syrinx, reported in association with Chiari I between 35 and 75% of pediatric patients. Similarly, in our series (440 females, 160 males, 98% > 18 years), syringomyelia associated with Chiari I was ranging from 40 to 60% (respectively in asymptomatic and symptomatic groups); headache was reported in 65%. Sensory disturbances (48%), cranial nerve deficits (45%), motor weakness (32%), and autonomic disorders (35%) were the most frequent neurological signs in our cohort. In Chiari I malformation, cervical pain and occipital cough headache are the most characteristic presenting symptoms, both in old children and in adults; however, headache is often multifactorial, and CM1 patients can report a wide variety of non-specific symptoms and signs. Clinical diagnostic CM1 criteria, shared at the national and international level, are recommended with the aim to avoid consequent controversies on diagnosis and on surgical decision making.

**Table 2** Neurological signs in patients with symptomatic Chiari I, Chiari Syndrome (CRESSC series)

| | All *n*=240 | Surgery (PFDD) *n*=142 | No Surgery *n*= 98 |
|---|---|---|---|
| **Sensory disorders (%)** [a] | **115 (48)** | **80 (56)** | **35 (36)** |
| Sensory Levels | 52 (22) | 27 (19) | 25 (26) |
| Hypoesthesia UL | 55 (23) | 30 (21) | 25 (26) |
| Hypoesthesia LL | 34 (14) | 30 (21) | 4 (4) |
| Paraesthesia/Dysesthesia | 19 (8) | 10 (7) | 9 (9) |
| Radicolar/Truncal deficit | 16 (6) | 3 (2) | 13 (13) |
| Dissociated Thermoalgesic loss | 10 (4) | 10 (7) | 0 |
| Deep sensitive disorders | 2 (1) | 1 (2) | 1 (1) |
| **Motor weakness (%)** [a] | **78 (32.5)** | **52 (36)** | **26 (26)** |
| Hyposthenia UL | 22 (9) | 17 (12) | 5 (5) |
| Upper MTN signs | 36 (15) | 31 (22) | 5 (5) |
| Lower MTN signs | 46 (19) | 30 (21) | 16 (16) |
| Hyposthenia LL | 15 (6) | 15 (11) | 0 |
| Spasticity | 18 (7.5) | 15 (11) | 3 (3) |
| Muscle atrophy | 12 (5) | 10 (7) | 2 (2) |
| Paraparesis | 12 (5) | 12 (8) | 0 |
| Tetraparesis | 10 (3) | 10 (7) | 0 |
| **Cranial Nerves disorders (%)** [a] | **109 (45)** | **77 (54)** | **32 (33)** |
| Dysphagia | 62 (26) | 40 (28) | 22 (22) |
| Nystagmus | 50 (21) | 43 (30) | 7 (7) |
| Dyspnea | 25 (10) | 25 (18) | 0 |
| Visual disorders | 15 (6) | 10 (7) | 5 (5) |
| Hearing loss | 9 (4) | 5 (3.5) | 4 (4) |
| **Sleep Disorders (%)** | **36 (15)** | **31 (22)** | **5 (5)** |
| OSAS | 24 (10) | 20 (14) | 4 (4) |
| CSAS | 12 (5) | 12 (8) | 0 |
| **Autonomic Disorders (%)** [a] | **74 (31)** | **70 (49)** | **4 (4)** |
| Incontinence | 22 (9) | 20 (14) | 2 (2) |
| Urge-incontinence | 52 (22) | 50 (35) | 2 (2) |
| Neurogenic bladder | 10 (4) | 10 (7) | 0 |
| **Gait Ataxia/cerebellar signs** | **18 (7.5)** | **13 (9)** | **5 (5)** |

[a] Patients who have at least one of neurological signs.

Abbreviations: UL-Upper Limbs; LL-Lower Limbs; MTN-Motoneurons; PFDD-Posterior Fossa Decompression and Duraplasty.

entrapment mononeuropathies (such as ulnar neuropathy at the elbow), were reported both in our cohort and in literature [17, 30]. In the chronic stage, upper motoneuron signs at the lower limbs could be also present, mimicking typical clinical manifestations of amyotrophic lateral sclerosis, another multilevel neuromuscular disease. Brainstem syndromes were frequently reported in Chiari syndrome and in siringobulbia, with nystagmus in 21–30% and dysphagia in 26–28%, similarly shown in previous series [21, 22]; central sleep apnea syndromes (CSAS) were evidenced in 5–8% by polysomnography, with the same frequency reported in a large pediatric study [8].

Neurophysiologie Clinique/Clinical Neurophysiology (2019) 49, 217–226

Disponible en ligne sur ScienceDirect www.sciencedirect.com

Elsevier Masson France EM|consulte www.em-consulte.com/en

ORIGINAL ARTICLE

# Brainstem evoked potentials and magnetic resonance imaging abnormalities in differential diagnosis of intracranial hypotension

Vincenzo Di Stefano[a,b,*], Camilla Ferrante[a,b], Roberta Telese[a,b], Massimo Caulo[a,c], Laura Bonanni[a,b], Marco Onofrj[a,b], Raffaella Franciotti[a,c]

[a] Department of Neuroscience, Imaging and Clinical Sciences, ''G. d'Annunzio'' University, Chieti, Italy
[b] Aging Research Centre, Ce.S.I., ''G. d'Annunzio'' University, Chieti, Italy
[c] Institute for Advanced Biomedical Technologies (ITAB), ''G. d'Annunzio'' University, Chieti, Italy

attenzione sulle forme Chiari-like da ipotensione intracranica, che possono associarsi a discesa tonsillare, sintomi cranio-cervicali e reperti neuroradiologici o neurofisiologici potenzialmente fuorvianti. Il punto, qui, non è tanto che questo studio spieghi direttamente la fisiopatologia della siringomielia, quanto che aiuti a evitare un errore concettuale preliminare: attribuire automaticamente un quadro con tonsillar descent a una Chiari primaria. In assenza di un corretto inquadramento anatomo-patogenetico, anche l'interpretazione dei dati neurofisiologici rischia infatti di diventare impropria.

*Conclusions.* — Our study points out how IH can be effectively distinguished from CM and SNHL through the contribution of neurophysiology and MRI; in particular, evaluation of the 8th nerve achieves a high sensitivity and specificity in patients with IH. Further studies are required to examine the combined use of BAEP recordings ad MRI in diagnosis and monitoring of patients affected by IH.

**Table 1** Symptoms, etiology, percentage of recovery due to treatment in IH patients.

| | % of patients |
|---|---|
| Symptoms | |
| Orthostatic headache | 100 |
| Chronic headache | 89 |
| Confusion | 22 |
| Lethargy | 22 |
| Tinnitus | 78 |
| Hearing loss | 78 |
| Hearing changes (alteration in the quality of sounds, i.e. metallic) | 56 |
| Neck stiffness | 56 |
| Etiology | |
| Trauma | 50 |
| Spontaneous (no apparent cause) | 33 |
| After epidural anesthesia | 11 |
| Marfan syndrome, with trauma | 6 |
| Treatment | |
| Bed rest (10—45 days) + hydration | 56 |
| Epidural blood patching (after 7—14 days) | 33 |

IH: intracranial hypotension. IH symptoms had been present for 48—750 days before diagnosis. In 12 patients, IH recovered spontaneously with bed rest and hydration in 10—45 days. Six patients were treated with epidural blood patching (and bed rest) and recovered in 7—14 days.

**Table 2** Auditory threshold (dB) in IH, CM and SNHL groups.

| | Auditory threshold (dB) | | |
|---|---|---|---|
| | 20—30 | 35—50 | > 50 |
| IH | 16 | 2 | None |
| CM | 18 | None | None |
| SNHL | 4 | 7 | 9 |

Auditory thresholds were derived from clicks delivered at a stimulus rate of 10 Hz.
IH: intracranial hypotension; CM: Chiari malformation; SNHL: sensorineural hearing loss.

**Table 3** Mean latencies of BAEP waves I, III and V in controls, IH, CM and SNHL patients. Test retest variability in controls and BAEP changes observed in IH after recovery.

| | I | II | III | V |
|---|---|---|---|---|
| Controls ($n=52$) | | | | |
| Mean latency ± SD | 1.58 ± 0.11 ($n=52$) | 2.47 ± 0.14 ($n=52$) | 3.59 ± 0.12 ($n=52$) | 5.61 ± 0.13 ($n=52$) |
| 3 SD upper latency limit | 1.91 | 2.89 | 3.95 | 6.00 |
| Test/retest latency change | ± 0.11 | ± 0.14 | ± 0.12 | ± 0.12 |
| IH ($n=18$) | | | | |
| Mean latency ± SD | 2.02 ± 0.13 ($n=15$) | 2.50 + 0.13 ($n=16$) | 3.72 ± 0.16 ($n=18$) | 5.93 ± 0.10 ($n=18$) |
| Delayed (D)/absent (A) % | D 77.8%/A 16.7% | D 5.5%/A 11.1% | D 16.7%/A None | D 27.8%/A None |
| IH post-recovery | | | | |
| Mean latency ± SD | 1.65 ± 0.11 ($n=18$) | 2.46 ± 0.14 ($n=18$) | 3.65 ± 0.12 ($n=18$) | 5.63 ± 0.12 ($n=18$) |
| Delayed (D)/absent (A) % | D None/A None | D None/A None | D None/A None | D None/A None |
| Latency changes (range) | −0.28 to 0.56 | +0.1 to 0.12 | +0.1 to 0.32 | −0.1 to 0.52 |
| CM ($n=18$) | | | | |
| Mean latency ± SD | 1.57 ± 0.11 ($n=16$) | 2.47 ± 0.12 ($n=18$) | 3.79 ± 0.12 ($n=18$) | 5.90 ± 0.08 ($n=18$) |
| Delayed (D)/absent (A) % | D None/A 11.1% | D None/A None | D 5.5%/A None | D 33.3%/A None |
| SNHL ($n=20$) | | | | |
| Mean latency ± SD | 1.49 ± 0.14 ($n=6$) | 2.45 ± 0.07 ($n=8$) | 3.58 ± 0.07 ($n=7$) | 5.92 ± 0.10 ($n=19$) |
| Delayed (D)/absent (A) % | D None/A 70% | D None/A 60% | D None/A 65% | D 20%/A 5% |

BAEP: brainstem acoustic evoked potentials. Waves were reported as delayed if the latencies were higher than the mean plus 3 standard deviations of the control groups. The latencies of the wave II were not different significantly among groups.
IH: intracranial hypotension; CM: Chiari malformation; SNHL: sensorineural hearing loss.

Neurophysiologie Clinique Clinical Neurophysiology

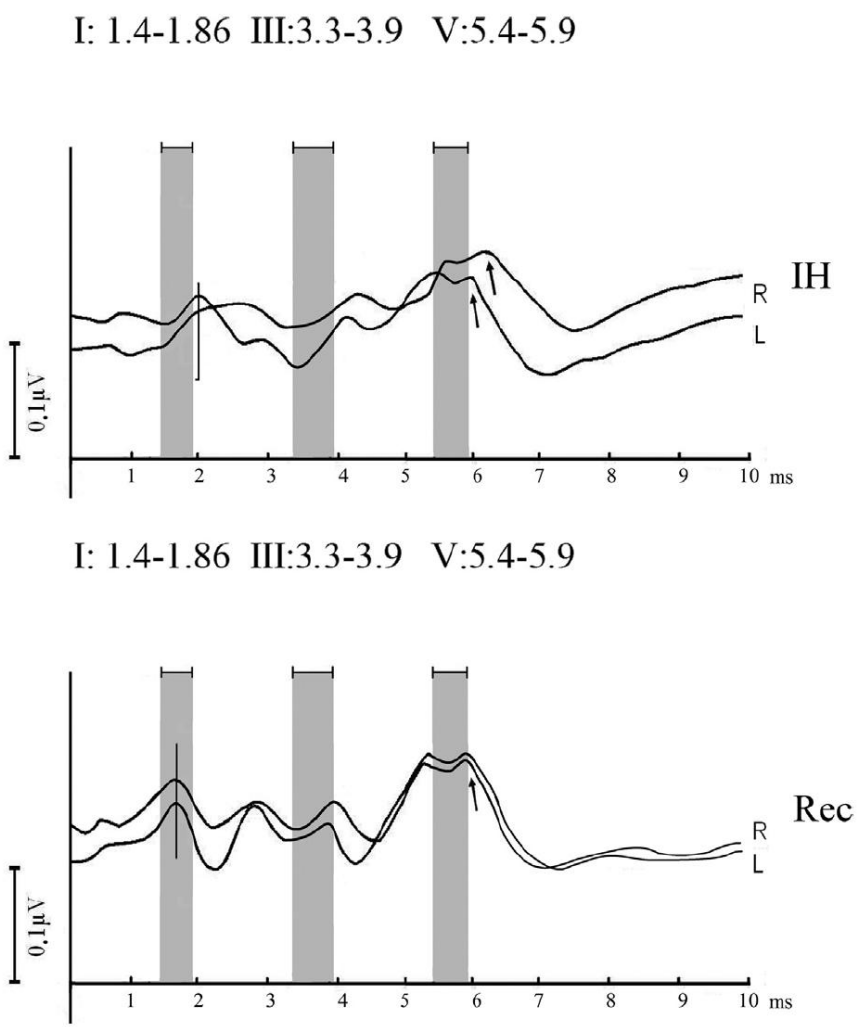



**Figure 1** Patterns of brainstem acoustic evoked potentials (BAEP) abnormalities in intracranial hypotension (IH). Vertical bands indicate latency ranges of components I, III and V in controls. Top: bilateral delayed wave I with delayed wave III—V during IH status (IH). Arrows point to delayed wave V. Bottom: latency delay reduction after recovery (Rec). Arrow points to normalized wave V latency. The vertical line marks wave I latency, which is in the normal range after recovery.

Neurophysiologie Clinique Clinical Neurophysiology



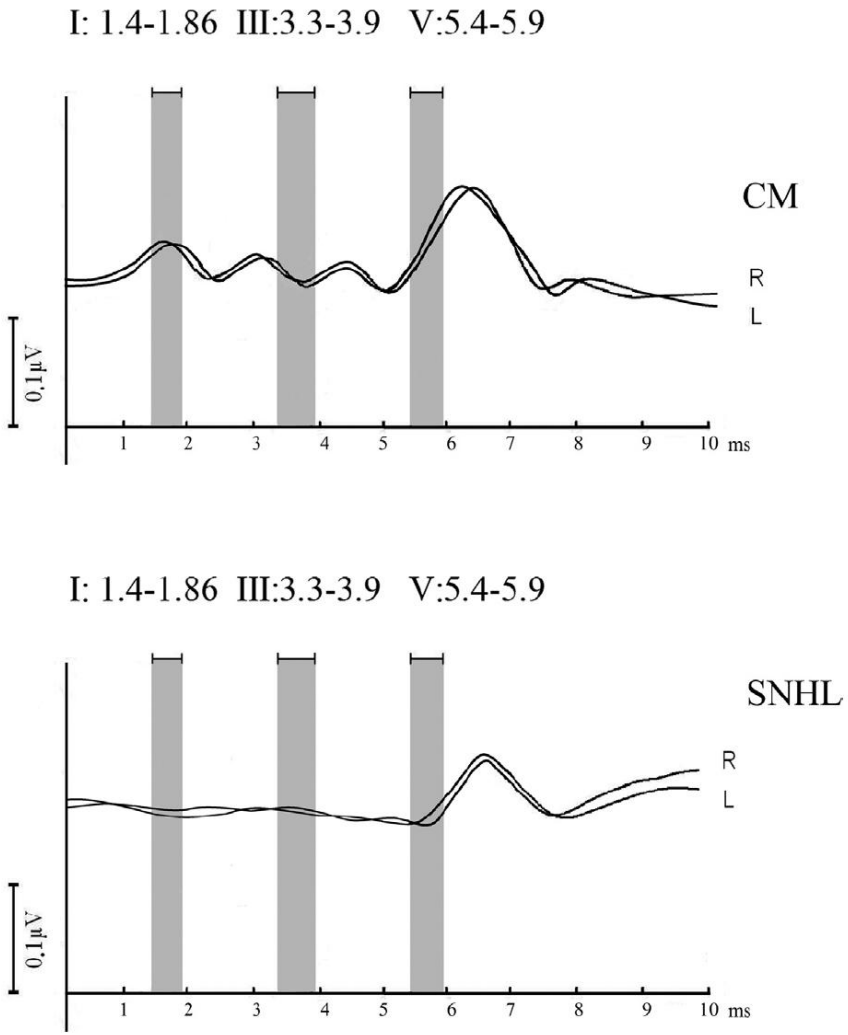



**Figure 2** Patterns of brainstem acoustic evoked potentials (BAEP) abnormalities in Chiari malformation (CM) and sensorineural hearing loss (SNHL). Top: bilateral delayed wave III and V with normal wave I in a patient affected by CM. Bottom: absent wave I and III, and delayed wave V in a SNHL patient. Vertical bars indicate latency ranges of component I, III, V in controls.

Journal of *Clinical Medicine*

MDPI

*Review*

# The Role of Neurophysiology in Managing Patients with Chiari Malformations

**Dulce Moncho [1,2,*], Maria A. Poca [2,3,4,*], Kimia Rahnama [1], M. Ángeles Sánchez Roldán [1], Daniela Santa-Cruz [1] and Juan Sahuquillo [2,3,4]**

1 Department of Clinical Neurophysiology, Vall d'Hebron University Hospital, Passeig Vall d'Hebron 119-129, 08035 Barcelona, Spain; kimia.rahnama@vallhebron.cat (K.R.); mariadelosangeles.sanchez@vallhebron.cat (M.Á.S.R.); danielaisabel.santacruz@vallhebron.cat (D.S.-C.)
2 Neurotraumatology and Neurosurgery Research Unit, Vall d'Hebron Institut de Recerca (VHIR), Vall d'Hebron Barcelona Hospital Campus, Passeig Vall d'Hebron 119-129, 08035 Barcelona, Spain; sahuquillo@neurotrauma.net
3 Department of Neurosurgery, Vall d'Hebron University Hospital, Passeig Vall d'Hebron 119-129, 08035 Barcelona, Spain
4 Department of Surgery, Universitat Autònoma de Barcelona, Bellaterra, 08193 Barcelona, Spain
* Correspondence: dulcemaria.moncho@vallhebron.cat (D.M.); pocama@neurotrauma.net (M.A.P.); Tel.: +34-93-274-62-41 (D.M.); Fax: +34-274-62-10 (D.M.)

**Abstract:** Chiari malformation type 1 (CM1) includes various congenital anomalies that share ectopia of the cerebellar tonsils lower than the foramen magnum, in some cases associated with syringomyelia or hydrocephalus. CM1 can cause dysfunction of the brainstem, spinal cord, and cranial nerves. This functional alteration of the nervous system can be detected by various modalities of neurophysiological tests, such as brainstem auditory evoked potentials, somatosensory evoked potentials, motor evoked potentials, electromyography and nerve conduction studies of the cranial nerves and spinal roots, as well as brainstem reflexes. The main goal of this study is to review the findings of multimodal neurophysiological examinations in published studies of patients with CM1 and their indication in the diagnosis, treatment, and follow-up of these patients, as well as their utility in intraoperative monitoring.

check for updates

Citation: Moncho, D.; Poca, M.A.;

**Table 3.** Summary of the characteristics of neurophysiological tests in CM1.

| Neurophysiological Tests | Pathway | Main Findings | Pitfalls and Limitations |
|---|---|---|---|
| SEP | Dorsal columns or lemniscal system | Increased N13-N20 interval. Reduced or absent cervical potential (N13) | - Associated neuropathy, other forms of cervical spinal stenosis, and cervical myelopathy<br>- Requires patient collaboration (especially relaxation) |
| BAEP | Auditory from cochlea to superior pons | Increased I–V interval, suggesting central or retrocochlear involvement | - Associated hypoacusia of cochlear origin<br>- Requires patient collaboration (especially relaxation) |
| MEP | Pyramidal system or corticospinal tract | Increased CMCT | - Variability related to maturative age<br>- Requires patient collaboration |
| EMG/NCS | Peripheral nervous system (roots/anterior horn, nerves and muscles) | Preganglionic pattern at NCS (preserved sensory neurography) with acute or chronic denervation at EMG, compatible with the suspicion of anterior horn cell lesion if syringomyelia is present | - Concomitant neuropathy or radiculopathy secondary to spondylosis |
| Brainstem reflexes (Blink reflex) | Involved cranial nerves, nucleus, and pathways in the brainstem | Altered R1 and/or R2 | - Variability related to maturative age<br>- Repetitive high frequencies stimuli cause adaptation phenomenon<br>- Lack of published follow-up studies for CM1 and other brainstem reflexes |
| Intraoperative neurophysiological monitoring | All of the pathways described above | Evidence for benefit during positioning | Anesthetic considerations |

BAEP: brainstem auditory evoked potential; CMCT: central motor conduction time; EMG: electromyography; MEP: motor evoked potential; NCS: nerve conduction studies; R: blink reflex response; SEP: somatosensory evoked potential.

La neurofisiologia aggiunge un' **informazione funzionale**. Non dice soltanto "dove" sia la lesione, ma aiuta a capire quali vie o circuiti siano già disfunzionanti, quali siano ancora risparmiati, e in quali pazienti il danno possa essere già presente anche se non ancora pienamente espresso clinicamente. È quindi utile non perché si sostituisca alla clinica o all'imaging, ma perché **consente di passare da una definizione anatomica della siringa a una definizione neurobiologica più precisa del caso**

# Brainstem Auditory and Somatosensory Evoked Potentials in Relation to Clinical and Neuroimaging Findings in Chiari Type 1 Malformation

*Dulce Moncho,*† Maria-Antonia Poca,†‡ Teresa Minoves,* Alejandro Ferré,*† Kimia Rahnama,* and Juan Sahuquillo†‡*

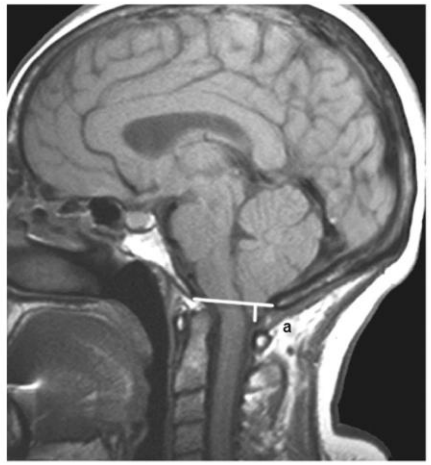



FIG. 1. Sagittal Midline T1-weighted MRI of a patient with Chiari type 1 malformation and a tonsillar descent of 5 mm below the foramen magnum (a).

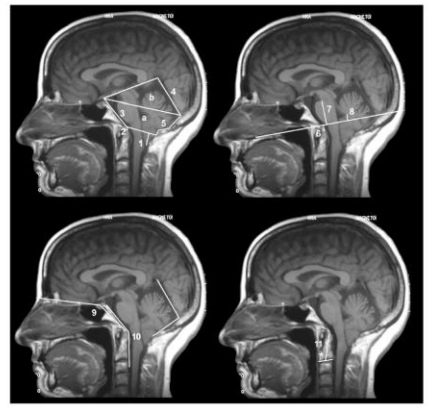


FIG. 2. Measurements of different anatomic structures of the posterior fossa (PF) and craniovertebral junction. 1, Descent (in millimeters) o the cerebellar tonsils below the foramen magnum. 2, The distance between the tip of the odontoid process and the Mc Rae line. 3, The length of the clivus. 4, The length of the tentorium. 5, The length of the suboccíput. 6, The distance between the tip of the odontoid process and the basal line. 7, The distance between the basal line and the pons. 8, The distance between the basal lin and the fastigium. 9, Basal angulation 10, The Wackenheim angle or clivus-canal angle. (a), the area limited by the PF bone contours. The sum of (a and (b) is the total area of the PF. 11* odontoid process angle.

I BAEP esplorano il funzionamento delle vie acustiche del tronco encefalico e, nel contesto di Chiari, possono risultare alterati soprattutto quando vi sia crowding della fossa posteriore, distorsione del tronco o interessamento della regione bulbopontina

I SSEP, invece, interrogano prevalentemente il sistema lemniscale e le colonne posteriori.
Nella siringomielia classica, soprattutto nelle fasi iniziali o nelle forme a prevalente coinvolgimento centrale, il sistema maggiormente interessato può essere quello spinotalamico;  di conseguenza, i SSEP possono anche essere poco alterati o normali, semplicemente perché non stanno esplorando il compartimento più colpito.
Quando invece la cavità si espande dorsalmente o il danno diventa più esteso, i SSEP tendono ad alterarsi più facilmente, perché vengono coinvolte anche le vie delle colonne posteriori. Questo spiega perché non sia corretto attribuire ai SSEP un valore “globale” sulla siringomielia: essi dicono molto su un sistema specifico, ma non sul significato funzionale della lesione

# Brainstem Auditory and Somatosensory Evoked Potentials in Relation to Clinical and Neuroimaging Findings in Chiari Type 1 Malformation

*Dulce Moncho,*† Maria-Antonia Poca,†‡ Teresa Minoves,* Alejandro Ferré,*† Kimia Rahnama,* and Juan Sahuquillo†‡*

**Purpose:** The aim of this study was to describe the abnormalities found in the recordings of evoked potentials (EPs), in particular those of brainstem auditory evoked potentials and somatosensory evoked potentials, in a homogeneous series of patients with Chiari type 1 malformation (CM-1) and study their relationship with clinical symptoms and malformation severity. CM-1 is characterized by cerebellar tonsils that descend below the foramen magnum and may be associated with EP alterations. However, only a small number of authors have described these tests in CM-1, and the patient groups studied to date have been small and heterogeneous.

**Methods:** The clinical findings, neuroimages, and EP findings were retrospectively studied in a cohort of 50 patients with CM-1.

**Results:** Seventy percent of patients had EP abnormalities (brainstem auditory evoked potential: 52%, posterior tibial nerve somatosensory evoked potential: 42%, and median nerve somatosensory evoked potential: 34%). The most frequent alteration was an increased central conduction time. Morphometric measurements differed between the normal and pathological groups, although no statistical significance was found when comparing these groups.

**Conclusions:** A high percentage of patients with CM-1 show EP alterations regardless of their clinical or radiological findings, thus highlighting the necessity of performing these tests, especially in patients with few or no symptoms.

**Key Words:** Brainstem auditory evoked potentials, Chiari type 1 malformation, Somatosensory evoked potentials, Syringomyelia, Study of the craniovertebral junction.

*(J Clin Neurophysiol* 2015;32: 130–138)

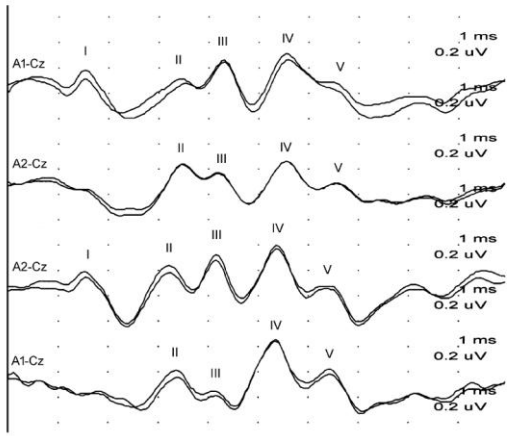



**FIG. 3.** Brainstem auditory evoked potentials in a 48-year-old woman with Chiari type 1 malformation. Tonsillar descent, 30 mm. The image shows the bilateral prolongation of latency of wave V (6.54 milliseconds, left ear; 6.40 milliseconds, right ear) and interpeak latency I–V (4.98 milliseconds, left ear; 4.86 milliseconds, right ear). The upper 2 channels record responses from stimulation of the left ear while the lower 2 channels represent stimulation of the right ear.

**TABLE 1.** Percentages of Altered Evoked Potentials in Patients With Chiari Type 1 Malformation

| | BAEP Unilateral (%) | BAEP Bilateral (%) | MN SEP Unilateral (%) | MN SEP Bilateral (%) | PTN SEP Unilateral (%) | PTN Bilateral (%) |
|---|---|---|---|---|---|---|
| Total (n = 50) | 22 (44) | 4 (8) | 12 (24) | 5 (10) | 14 (28) | 7 (14) |
| Without syringomyelia (n = 30) | 12 (40) | 3 (10) | 7 (23.3) | 3 (10) | 9 (30) | 3 (10) |
| With syringomyelia (n = 20)* | 10 (50) | 1 (5) | 5 (25) | 2 (10) | 5 (25) | 4 (20) |
| With retroflexed odontoid (n = 9)† | 4 (44.4) | 1 (11.1) | 2 (22.2) | 0 (0) | 3 (33.3) | 0 (0) |

*Absence of the N28 potential with cortical potential P37/N45 alterations observed in the three cases of severe syringomyelia.
†Two patients had simultaneous alterations in the three types of EPs.
BAEP, brainstem auditory evoked potentials; MN SEP, median nerve somatosensory evoked potentials; PTN SEP, posterior tibial nerve somatosensory evoked potentials.

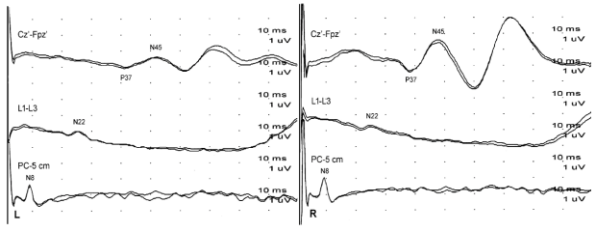



**FIG. 4.** Posterior tibial nerve somatosensory evoked potentials in a 27-year-old man with Chiari type 1 malformation. Tonsillar descent, 18.7 mm. Syringomyelia between the vertebral bodies C4 and D2. Height, 178 cm. The image shows an asymmetry of cortical potential, with a more prolonged N22-P37 interpeak latency (16.6 milliseconds vs 13.6 milliseconds) and a lower amplitude (0.675 μV vs 2.030 μV) on the left when compared with the right. PC, popliteal crease. R, right; L, left.

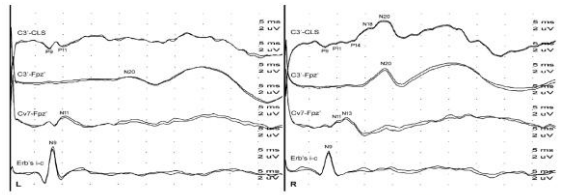



**FIG. 5.** Median nerve evoked potentials in a 63-year-old woman with Chiari type 1 malformation. Tonsillar descent, 10.6 mm. No syringomyelia. Height, 150 cm. The image shows a clear asymmetry, with an absence of cervical potential and poorly defined, smaller amplitude, as well as a prolonged latency of the cortical potential on the left. What is also remarkable is a N13-N20 interpeak latency that is prolonged (7 milliseconds) on the right side. CLS, contralateral shoulder; Cv7, seventh cervical vertebra; R, right; L, left.

**TABLE 2.** Alterations Detected in the Different Parameters Recorded in EPs

| | Unilateral Prolonged Latency | Bilateral Prolonged Latency | Reduced Amplitude | Absence |
|---|---|---|---|---|
| BAEPs | | | | |
| Wave I | 0 | 0 | — | 0 |
| Wave III | 8 (16%) | 0 | — | 2 (4%) |
| Wave V | 21 (42%) | 3 (6%) | — | 0 |
| I–III IPL | 7 (14%) | 2 (4%) | — | 2 (4%) |
| III–V IPL | 8 (16%) | 1 (2%) | — | 2 (4%) |
| I–V IPL | 21 (42%) | 3 (6%) | — | 0 |
| PTN SEPs | | | | |
| N8 | 0 | 0 | 0 | 0 |
| N22 | 0 | 0 | 0 | 0 |
| N28 | 1 (2%) | 1 (2%) | 0 | 8 (16%) |
| P37/N45 | 13 (26%) | 6 (12%) | 7 (14%) | 0 |
| N22–P37 IPL | 13 (26%) | 6 (12%) | — | 0 |
| MN SEPs | | | | |
| Wave N9 | 0 | 0 | 0 | 0 |
| Wave N13 | 2 (4%) | 0 | 1 (2%) | 1(2%) |
| Wave N20 | 4 (8%) | 1 (2%) | 6 (12%) | 0 |
| N9–N13 IPL | 1 (2%) | 0 | — | 1(2%) |
| N13–N20 IPL | 7 (14%) | 5 (10%) | — | 1(2%) |

BAEPs, brainstem auditory evoked potentials; IPL, interpeak latency; PTN SEPs, posterior tibial nerve somatosensory evoked potentials; MN SEPs, median nerve somatosensory evoked potentials.

**TABLE 3.** Summary of the Most Relevant Comparative Statistical Studies Assessed

| Morphological Measurement | Patients With Normal BAEP (n = 24) | Patients with Pathological BAEP* (n = 26) | U | P |
|---|---|---|---|---|
| Tonsillar descent below FM, mm | n = 24<br>Median: 7.0<br>IQR: 3.8 | n = 25<br>Median: 11.0<br>IQR: 12.0 | 231.5 | 0.173 |
| Tonsillar descent below basal line, mm | n = 24<br>Median: 6.0<br>IQR: 9.8 | n = 24<br>Median: 10.6<br>IQR: 12.0 | 205.0 | 0.088 |
| **Morphological Measurement** | **Patients With Normal PTN SEP (n = 29)** | **Patients With Pathological PTN SEP† (n = 21)** | **U** | **P** |
| Tonsillar descent below FM, mm | n = 28<br>Median: 7.0<br>IQR: 7.7 | n = 21<br>Median: 10.0<br>IQR: 7.0 | 200.0 | 0.058 |
| Tonsillar descent below basal line, mm | n = 28<br>Median: 6.0<br>IQR: 11.6 | n = 20<br>Median: 11.6<br>IQR: 8.5 | 193.0 | 0.070 |
| **Morphological Measurement** | **Patients With Normal MN SEP (n = 33)** | **Patients With Pathological MN SEP‡ (n = 17)** | **U** | **P** |
| Tonsillar descent below FM, mm | n = 32<br>Median: 7.0<br>IQR: 7.7 | n = 17<br>Median: 11.0<br>IQR: 7.5 | 201.5 | 0.141 |
| Distance between basal line and fastigium, mm | n = 32<br>Median: 26.0<br>IQR: 6.8 | n = 16<br>Median: 29.1<br>IQR: 6.1 | 167.0 | 0.052 |

Comparison of the morphological parameters of the PF between patients with normal and pathological EPs (Mann–Whitney Rank-Sum Test).
*Includes 21 patients with a unilateral alteration and 5 with a bilateral alteration in BAEP recordings.
†Includes 14 patients with a unilateral alteration and 7 with a bilateral alteration in PTN SEP recordings.
‡Includes 12 patients with a unilateral alteration and 5 with a bilateral alteration in MN SEP recordings.
BAEPs, brainstem auditory evoked potentials; EPs, evoked potentials; FM, foramen magnum; IQR, interquartile range; MN SEPs, median nerve somatosensory evoked potentials; PF, posterior fossa; PTN SEPs, posterior tibial nerve somatosensory evoked potentials.

# Are evoked potentials clinically useful in the study of patients with Chiari malformation Type 1?

Dulce Moncho, MD,[1,3] Maria A. Poca, MD, PhD,[2,3] Teresa Minoves, MD,[1] Alejandro Ferré, MD,[1,3] Victoria Cañas, MSc,[3] and Juan Sahuquillo, MD, PhD[2,3]

[1]Clinical Neurophysiology Department, [2]Neurosurgical Department, and [3]Neurotraumatology and Neurosurgery Research Unit (UNINN), Vall d'Hebron University Hospital, Vall d'Hebron Research Institute, Universitat Autònoma de Barcelona, Spain

**TABLE 3. Evoked potential findings in the different variants of CM***

| EPs | CM-0 | | CM-1 | | CM-1.5 | | Total Cohort | |
|---|---|---|---|---|---|---|---|---|
| | Yes (n = 9) | No (n = 5) | Yes (n = 70) | No (n = 67) | Yes (n = 17) | No (n = 32) | Yes (n = 96) | No (n = 104) |
| Normal BAEPs/SSEPs (%) | 3 (33.3) | 3 (60.0) | 24 (34.3) | 32 (47.8) | 7 (41.2) | 11 (34.4) | 34 (35.4) | 46 (44.2) |
| Abnormal BAEPs/SSEPs (%) | 2 (22.2) | 2 (40.0) | 16 (22.9) | 10 (14.9) | 4 (23.5) | 10 (31.3) | 22 (22.9) | 22 (21.2) |
| Normal BAEPs & abnormal SSEPs (%)† | 4 (44.4) | 0 (0.0) | 20 (28.6) | 12 (17.9) | 4 (23.5) | 3 (9.4) | 28 (29.2) | 15 (14.4) |
| Abnormal BAEPs & normal SSEPs (%) | 0 (0.0) | 0 (0.0) | 10 (14.3) | 13 (19.4) | 2 (11.8) | 8 (25.0) | 12 (12.5) | 21 (20.2) |

* Yes/no refers to the presence/absence of syringomyelia.

† Normal BAEPs and abnormal SSEPs were more frequent in patients with syringomyelia ($\chi^2 = 6.43$, df = 1, p = 0.011).

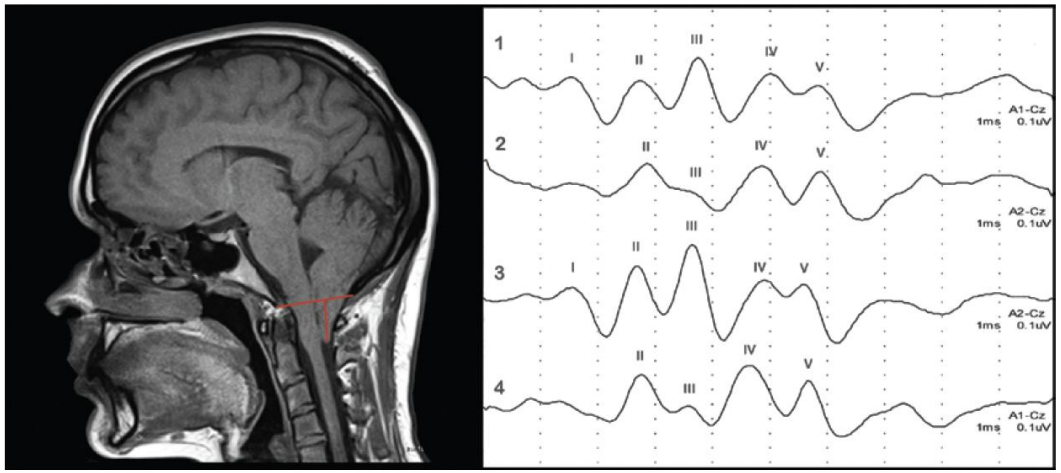



**FIG. 2.** BAEPs in a 26-year-old woman with CM-1.5. The sagittal midline T1-weighted MR image (**left**) shows a 20-mm tonsillar descent below the FM. Patient symptoms include headaches in an occipitonuchal location, exacerbated with Valsalva maneuvers. The graph (**right**) shows responses obtained from stimulation of the left ear (channels 1 and 2) and responses obtained from stimulation of the right ear (channels 3 and 4). BAEPs show an asymmetrical IPL I–V, longer in the left (4.29 msec) than in the right ear (3.98 msec). Figure is available in color online only.

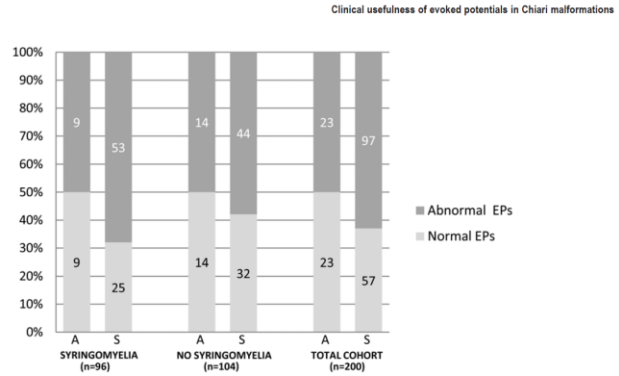



**FIG. 3.** Stacked bar showing the percentages of abnormal and normal EPs in asymptomatic (A) and symptomatic (S) patients. Both groups with and without syringomyelia, as well in the total cohort, are shown. The number of patients is shown inside the stacked bars.

BAEP e SSEP si alterano spesso, ma la loro utilità maggiore non è nei pazienti già chiaramente sintomatici, dove la clinica e l'imaging sono spesso sufficienti, bensì nei casi incidentali, asintomatici o paucisintomatici, nei quali possono documentare una sofferenza subclinica e contribuire al follow-up

In altre parole, gli evocati non sono tanto "test di indicazione chirurgica", quanto strumenti di staging funzionale

Questo chiarisce anche perché, sul piano fisiopatologico, metodiche nocicettive come i laser evoked potentials conservino un interesse teorico particolare. Se la siringa colpisce preferenzialmente le fibre spinotalamiche crociate, è intuitivo che una metodica centrata sulle vie nocicettive sia più "congruente" con il danno atteso rispetto ai SSEP. Il fatto che tali tecniche siano meno diffuse nella pratica non ne annulla il valore concettuale: anzi, conferma quanto sia importante scegliere il test in base alla fisiologia del sistema che si vuole esplorare-

*Brain* (1991), **114**, 1595–1607

# CEREBRAL POTENTIALS EVOKED BY PAINFUL LASER STIMULI IN PATIENTS WITH SYRINGOMYELIA

*by* R.-D. TREEDE,[1] J. LANKERS,[1] A. FRIELING,[1] W. H. ZANGEMEISTER,[2] K. KUNZE[2] *and* B. BROMM[1]

*(From the [1]Institute of Physiology and [2]Department of Neurology, University Hospital Eppendorf, D-2000 Hamburg 20, Germany)*

## SUMMARY

Brief cutaneous heat stimuli generated by a $CO_2$ laser were used to elicit late somatosensory evoked cerebral potentials (SEPc) in 10 patients with syringomyelia. For comparison, early and late cerebral potentials in response to electrical nerve stimuli (SEPn) were recorded in the same session. In 8 patients with localized impairment of pain and temperature sensitivity we found complete absence of SEPc after stimulation of the affected area; in another patient with similar sensory deficits, the SEPc was grossly attenuated and delayed. In 1 patient with intact pain sensitivity but absent temperature sensitivity, a well defined SEPc could be recorded. Both early cortical SEPn and late SEPn in response to conventional nerve stimuli were normal in all patients and thus did not differentiate control and affected areas. These data indicate that alteration of SEPc correlates with altered pain sensitivity in patients with a circumscribed spinal lesion. SEPc may thus be used as a neurophysiological test in the assessment of hypalgesic dermatomes.

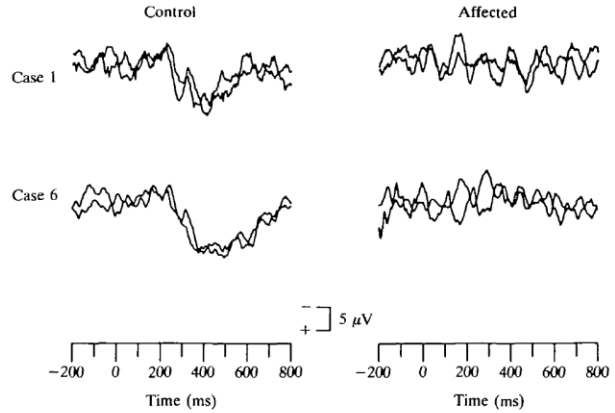



FIG. 1. Late somatosensory evoked potentials in response to cutaneous heat stimulation (SEPc) of the affected left hand and the unaffected right hand in 2 patients with syringomyelia. Each trace represents an average across 20 repetitions of the 20 W stimulus. Vertex versus linked earlobes, negativity upwards. *Top row: Case 1*, a man aged 47 y with a cervical syrinx. Pain and temperature sensitivity were lost over the left upper body quadrant including the left face. *Bottom row: Case 6*, a man aged 45 y with a cervicothoracic syrinx and Chiari I malformation. He had complete analgesia over the left upper body quadrant without affection of the face. In both cases, the SEPc were normal after stimulation of the control site and absent after stimulation of the affected site.



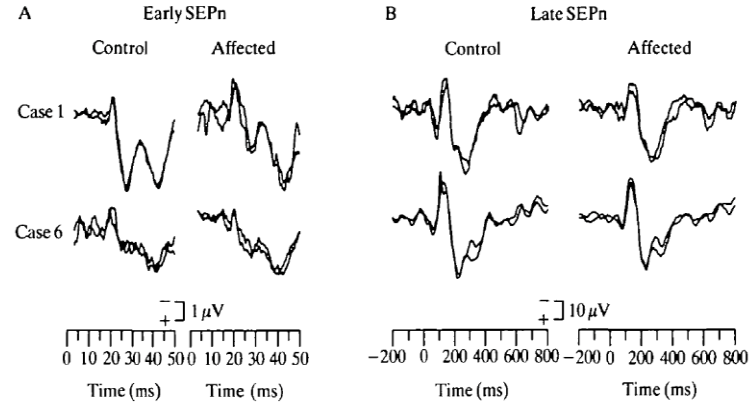



FIG. 2. Somatosensory evoked potentials in response to electrical stimulation of the median nerve (SEPn) of the affected left hand and the unaffected right hand in 2 patients with syringomyelia. Same patients as in fig. 1 (*top and bottom*). A, early SEPn components. Each trace represents an average across 256 stimulus repetitions. Hand projection area (2 cm posterior to C3 or C4) versus Fz, negativity upwards. B, late SEPn components. Each trace represents an average across 20 stimulus repetitions. Vertex versus linked earlobes, negativity upwards. In both cases, all SEPn were normal after stimulation of both test sites.

TABLE 3. NEUROPHYSIOLOGICAL DATA

| | *Late SEPc Peak-to-peak amplitudes (μV)* | | *Late SEPn Peak-to-peak amplitudes (μV)* | | *Early SEPn Latencies (ms)* | | | |
|---|---|---|---|---|---|---|---|---|
| | | | | | *Control* | | *Affected* | |
| *Case* | *Control* | *Affected* | *Control* | *Affected* | *Med.n* | *Tib.n* | *Med.n* | *Tib.n* |
| 1 | 14 | – | 47 | 39 | 21 | n.d. | 20 | n.d. |
| 2 | 12 | – | 12 | 26 | n.d. | 49* | 21 | n.d. |
| 3 | 12 | – | 26 | 30 | n.d. | 43 | 21 | n.d. |
| 4 | 56 | 10* | 69 | 37 | n.d. | – | 19 | n.d. |
| 5 | 30 | 36 | 58 | 60 | 22 | n.d. | n.d. | 46 |
| 6 | 14 | – | 51 | 47 | 20 | n.d. | 20 | n.d. |
| 7 | – | – | 37 | 52 | 21 | n.d. | 22 | n.d. |
| 8 | 34 | – | 35 | 28 | 22 | n.d. | 22 | n.d. |
| 9 | 5 | – | 15 | 13 | n.d. | 45 | 22 | n.d. |
| 10 | 16 | – | 33 | 62 | – | n.d | 22 | n.d. |

– = absent response; * = delayed response; n.d. = not determined; Med.n = median nerve; Tib.n = tibial nerve.

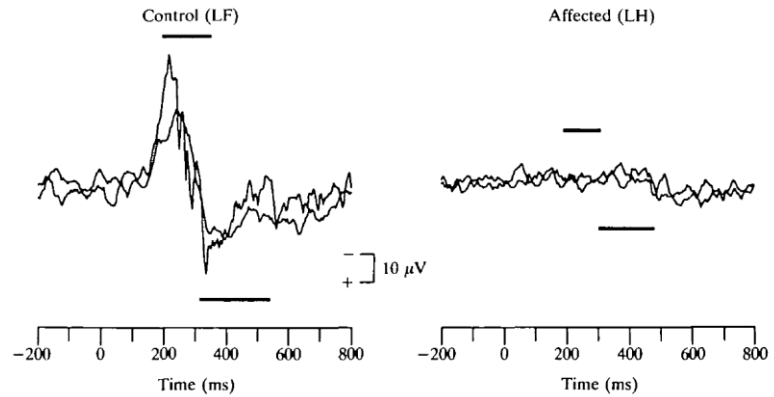



FIG. 3. Late somatosensory evoked potentials in response to cutaneous heat stimulation (SEPc) of the affected left hand (LH) and the unaffected left foot (LF) in a woman aged 50 y with a thoracocervical syringomyelia (*Case 4*). Each trace represents an average across 20 stimulus repetitions. Vertex versus linked earlobes, negativity upwards. Pain and temperature sensitivity was lost bilaterally from the trigeminal region down to T4 dermatome. The SEPc for the affected site was greatly attenuated, and its latency was outside the 3 SD range in normal subjects (horizontal bars).



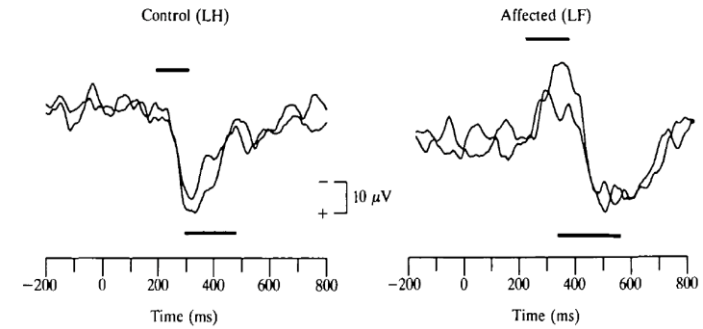



FIG. 4. Late somatosensory evoked potentials in response to cutaneous heat stimulation (SEPc) of the affected left foot (LF) and the unaffected left hand (LH) in a man aged 18 y with cervical syringomyelia due to a central tumour at the C3–C4 level (*Case 5*). Each trace represents an average across 20 stimulus repetitions. Vertex versus linked earlobes, negativity upwards. The horizontal bars indicate the range of latency variability (±3 SD) of the respective SEPc components in normal subjects. Temperature sensitivity was lost at both feet while pain sensitivity was not disturbed. In this case SEPc were normal in all investigated areas.

SPINE Volume 34, Number 14, pp E484–E487
©2009, Lippincott Williams & Wilkins

# Thoracic Disc Herniation in a Patient With Tethered Cord and Lumbar Syringomyelia and Diastematomyelia

## Magnetic Resonance Imaging and Neurophysiological Findings

John L. K. Kramer, MSc,*† Marcel Dvorak, MD,*‡ and Armin Curt, MD*‡

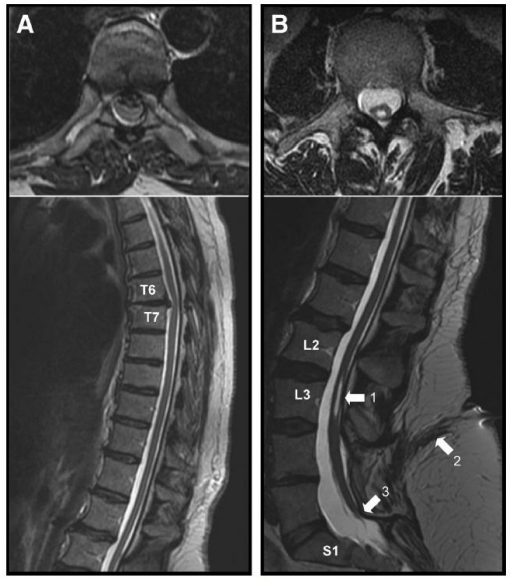



Figure 1. **A,** Axial and sagittal MRI of T6–T7 disc herniation. **B,** Axial and sagittal MRI L2–L3 syrinx (1). Closed spina bifida (2) and tethered cord (3) are also evident in the sagittal image.

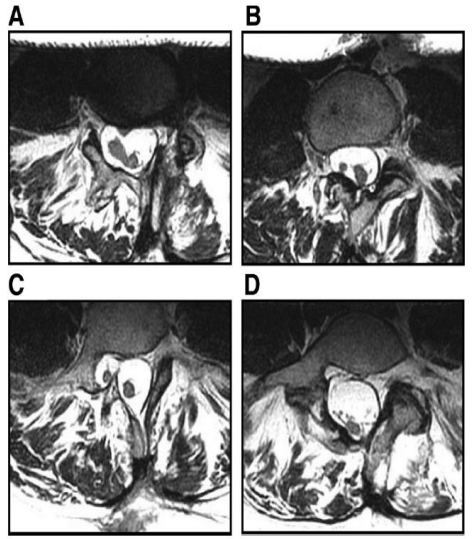



Figure 2. Axial MRI slices of the lumbar spine. **A,** Lumbar segment rostral to diastematomyelia, (**B, C**) lumbar segments at diastematomelia, and (**D**) lumbar segment and cauda equina caudal to diastematomyelia.

la combinazione di RM, SSEP ed EMG consente di distinguere la lesione semplicemente visibile da quella realmente responsabile del quadro clinico. È un punto molto rilevante, perché nei pazienti con anatomia complessa o con più reperti concomitanti la domanda clinicamente importante non è solo “quali lesioni ci sono?”, ma “quale di queste spiega davvero i sintomi del paziente?”. La neurofisiologia, in questi casi, può agire come strumento di localizzazione funzionale, e quindi avere un impatto molto concreto sulla strategia terapeutica



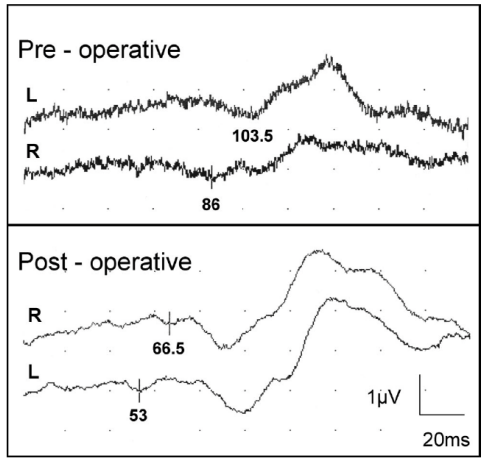



Figure 3. Pre- and post T6–T7 decompression tibial somatosensory-evoked potentials. Normal peripheral nerve conduction recorded at the popliteal fossa (not shown) was observed bilaterally. All values reflect millisecond. L, left; R, right.

### Key Points

- Neurophysiological testing can assist in determining the clinical relevance of spinal pathologies disclosed by neuroimaging.
- Diagnosing a symptomatic TDH can be challenging, not only in patients with additional lumbar cord abnormalities.
- Coupling segmental nerve conduction studies and EMG with longitudinal SSEPs is clinically useful for dissociating multiple levels of spinal cord pathology.

ABSTRACT: Electrodiagnostic abnormalities are well known to occur in syringomyelia although the findings are nonspecific. The objective of this work was to describe different types of spontaneous electromyographic (EMG) activity and reflex responses, which may be useful and more specific than conventional findings for the electrodiagnosis of syringomyelia. We studied 43 patients with syringomyelia by four-channel surface EMG and by recording the long-latency responses to distal stimulation of the median and tibial nerves. Continuous motor unit activity (CMUA) was found in 18 patients, synchronous motor unit potentials (SMUP) in 10, respiratory synkinesis (RS) in 5, and myokymic discharges in 4. Long-latency responses (LLR) with latencies ranging from 55 to 150 ms were found in 14 patients. Patients with syringomyelia thus show a wide variation of spontaneous EMG activity. An increase in excitability of spinal motor neurons is probably the basic underlying mechanism.
© 1999 John Wiley & Sons, Inc. *Muscle Nerve* **22:** 1653–1659, 1999

# ELECTRODIAGNOSTIC FINDINGS IN SYRINGOMYELIA

**MARTÍN A. NOGUÉS, MD,[1] and ERIK STÅLBERG, MD[2]**

[1] Department of Clinical Neurophysiology, Raúl Carrea Institute for Neurological Research (FLENI), Montañeses 2325, 1428 Buenos Aires, Argentina
[2] Department of Clinical Neurophysiology, University Hospital, Uppsala, Sweden

*Accepted 10 July 1999*

**Synchronous Motor Unit Potentials.** SMUPs were observed in 8 patients in the finger flexors and extensor muscles (Fig. 1), and in 2 others in tibialis anterior and gastrocnemius muscles bilaterally. SMUPs were either simple or complex motor unit potentials firing irregularly and continuously in two antagonistic limb muscles, either finger flexors and extensors, or tibialis anterior and gastrocnemius

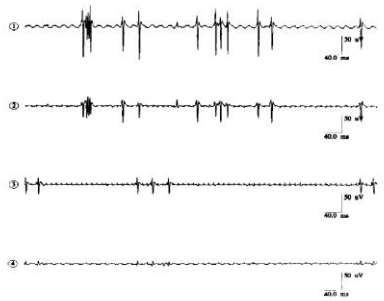


**FIGURE 1.** CMUA and SMUPs recorded from antagonistic forearm muscles at rest. Simultaneous surface EMG recording from left finger flexors (channel 1), left finger extensors (channel 2), right finger flexors (channel 3), and right finger extensors (channel 4). There is synchronicity between MUPs in channels 1–2 and 3–4, respectively. A single burst of MUPs is seen early in traces 1 and 2 (the more affected limb), associated with finger flexion movements (minipolymyoclonus).

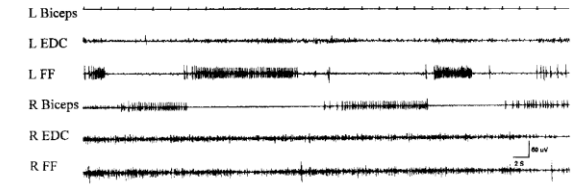



**FIGURE 2.** Myokymic discharges (surface recording). Long-duration rhythmic and "alternating" myokymic discharges in left fingers flexors and contralateral biceps brachii muscles. CMUA also occurs in right finger flexors and extensor digitorum communis muscles. L: left, R: right; EDC: extensor digitorum communis; FF: finger flexors.

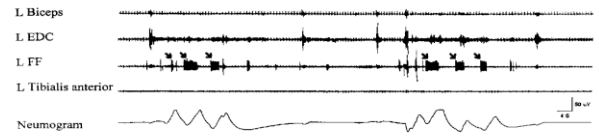



**FIGURE 3.** Respiratory synkinesis ("breathing arm") (surface recording). Periodic breathing in a patient with syringomyelia and syringobulbia. Each inspiration is associated with a MUP burst in the left finger flexors (arrows).

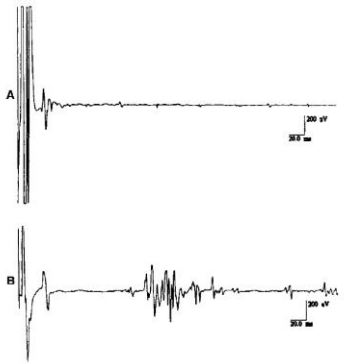



**FIGURE 4. (A)** CMAP and F-wave response after supramaximal electrical median nerve stimulation at the wrist in a normal subject. **(B)** Response to the same stimuli in a patient with a cervical syrinx and a Chiari anomaly. CMAP, F response, and LLR with a latency of 160 ms.

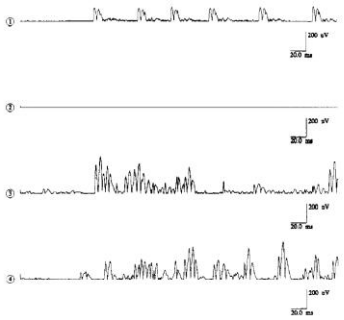


**FIGURE 5.** LLR to supramaximal left tibial nerve stimulation at the ankle. Single stimulus at 60 mA with a duration of 0.5 s (surface recording). Channel 1: right biceps brachii; channel 2: right extensor digitorum communis; channel 3: left biceps brachii; channel 4: left extensor digitorum communis. Stimulation evokes a brief arm jerk (flexion at the elbow). A long sweep speed is used to show the duration of the response and its repetitiveness (most clearly seen in channel 1). Signals are rectified.

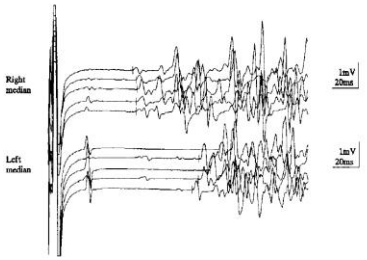



**FIGURE 6.** Upper panel: Mixed nerve silent period elicited by right (affected side) median nerve stimulation at the wrist during voluntary activation of the right abductor pollicis brevis (APB) muscle in a patient with dissociated sensory loss restricted to the right arm and upper trunk. Lower panel: Mixed nerve silent period evoked by stimulation of the left median nerve at the wrist and recording from the APB muscle in the same patient. Note that the duration of the silent period on the left (healthy) side is approximately double that of the affected side (surface recording).

l'EMG nella siringomielia non coincide con un semplice pattern neurogeno cronico. La presenza di continuous motor unit activity, synchronous motor unit potentials, respiratory synkinesis, scariche miochimiche e long-latency responses suggerisce che il midollo colpito dalla siringa non sia solo "leso", ma anche funzionalmente disinibito . Questo è il punto neurofisiologico più importante: **l'interessamento della sostanza grigia centrale e dei circuiti segmentari può produrre una perdita del controllo inibitorio locale, con conseguente comparsa di ipereccitabilità spinale** .

L'EMG, quindi, non documenta solo la perdita di motoneuroni segmentari, ma anche la **disorganizzazione del network che normalmente regola la scarica motoria a quel livello**

ABSTRACT: Different types of spontaneous activity may be found d
electromyographic examinations in patients with spinal cord diseases
ringomyelia and intramedullary tumor patients may show continuous r
unit activity, synchronous motor unit potentials, myokymic discharges,
mental and propriospinal myoclonus, and respiratory synkinesis. T
types of discharges are less commonly encountered in other types of s
cord lesions. It is suggested that the derangement of inhibitory mechar
by a central spinal cord lesion may favor the appearance of abnormal s
taneous activity. An increase in the excitability of spinal motor neuro
probably the basic underlying mechanism.
© 2002 Wiley Periodicals, Inc. *Muscle Nerve Supplement* **11:** S77–S82,

# SPONTANEOUS ELECTROMYOGRAPHIC ACTIVITY IN SPINAL CORD LESIONS

**MARTÍN A. NOGUÉS, MD**

Department of Clinical Neurophysiology, Instituto de Investigaciones Neurológicas "Dr. Raúl Carrea" (FLENI), Montañeses 2325, 1428 Buenos Aires, Argentina

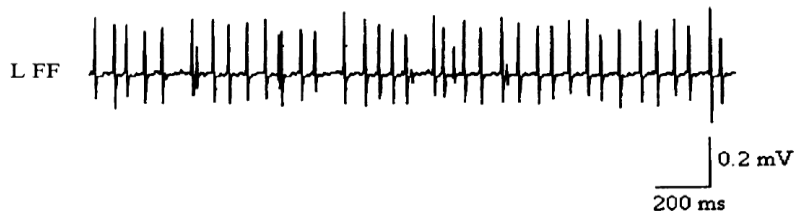



**FIGURE 1.** CMUA recorded from the left finger flexor (FF) muscles in a patient with syringomyelia (concentric needle recording).

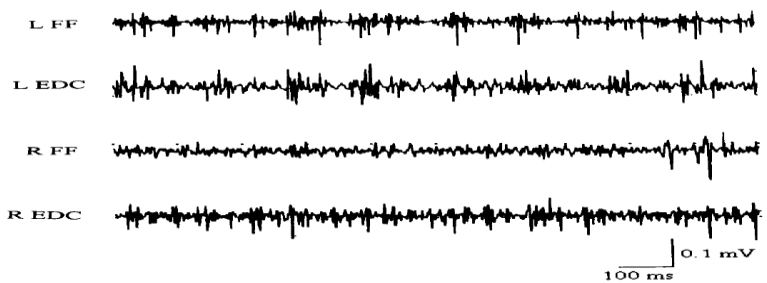



**FIGURE 2.** CMUP and SMUP recorded from the left and right finger flexor (FF) and extensor digitorum communis (EDC) muscles. Despite voluntary relaxation of the limb, there is spontaneous firing of MUPs in forearm muscles. Some MUPs are synchronous in antagonist muscles of the left forearm (left FF and EDC).

**Table 1.** Spontaneous EMG activity that may be recorded in patients with spinal cord lesions.

| |
|---|
| Spinal origin (montoneurons, interneurons, and propriospinal pathways) |
| Fasciculations |
| Myokymia |
| CMUA |
| SMUPs |
| RS |
| Myoclonus, segmental and propriospinal |
| Tremor |
| Prejunctional (roots and motor axons) |
| Fasciculations |
| Myokymia |
| RS |
| Cramps |
| Postjunctional (denervated muscle) |
| Fibrillations |
| Positive sharp waves |
| Complex repetitive discharges |

ABSTRACT: Different types of spontaneous activity may be found c
electromyographic examinations in patients with spinal cord diseases
ringomyelia and intramedullary tumor patients may show continuous r
unit activity, synchronous motor unit potentials, myokymic discharges,
mental and propriospinal myoclonus, and respiratory synkinesis. T
types of discharges are less commonly encountered in other types of s
cord lesions. It is suggested that the derangement of inhibitory mechar
by a central spinal cord lesion may favor the appearance of abnormal s
taneous activity. An increase in the excitability of spinal motor neuro
probably the basic underlying mechanism.
© 2002 Wiley Periodicals, Inc. *Muscle Nerve Supplement* **11:** S77–S82,

# SPONTANEOUS ELECTROMYOGRAPHIC ACTIVITY IN SPINAL CORD LESIONS

**MARTÍN A. NOGUÉS, MD**

Department of Clinical Neurophysiology, Instituto de Investigaciones Neurológicas "Dr. Raúl Carrea" (FLENI), Montañeses 2325, 1428 Buenos Aires, Argentina

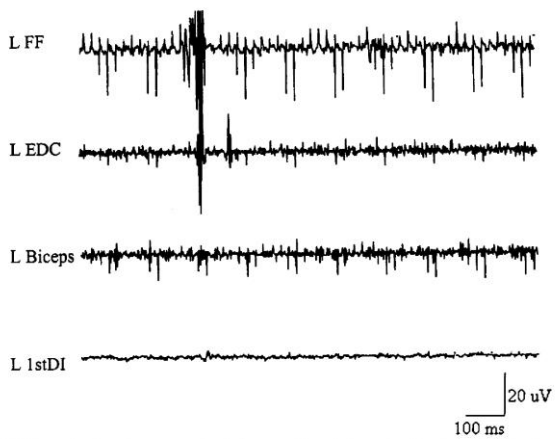



**FIGURE 4.** Surface EMG recording from forearm muscles during complete voluntary relaxation. A myoclonic burst associated with a brief jerk (flexion left hand and fingers) is recorded from the left finger flexor (FF) and extensor digitorum communis (EDC) muscles. Spontaneous MUPs are also recorded as background activity from the left FF, EDC, and biceps brachii muscles.

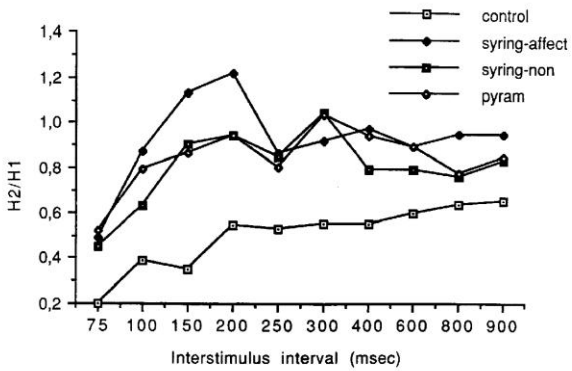



**FIGURE 5.** Integrated H-reflex recovery curves in normal subjects (control, *n* = 12), patients with upper motor neuron weakness due to cerebrovascular disease (pyram, *n* = 8), and syringomyelia patients (syring-affect = more affected limb, syring-non = less affected limb, *n* = 13). Recovery is maximal with interstimulus intervals of 150–200 ms.

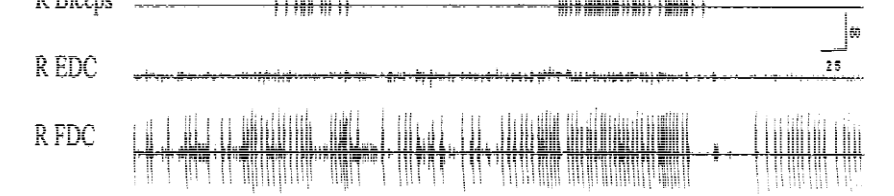



**FIGURE 3.** Myokymic discharges: long-duration rhythmic and alternating myokymic discharges in left flexor digitorum communis (FDC) and contralateral biceps brachii muscles. CMUA is also recorded from the right FDC and extensor digitorum communis (EDC) muscles (surface recording).

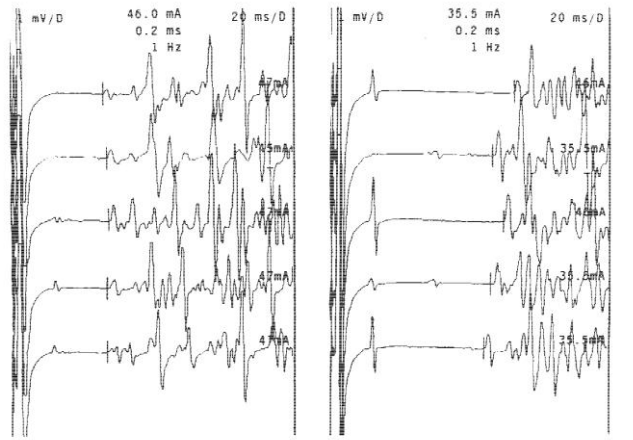
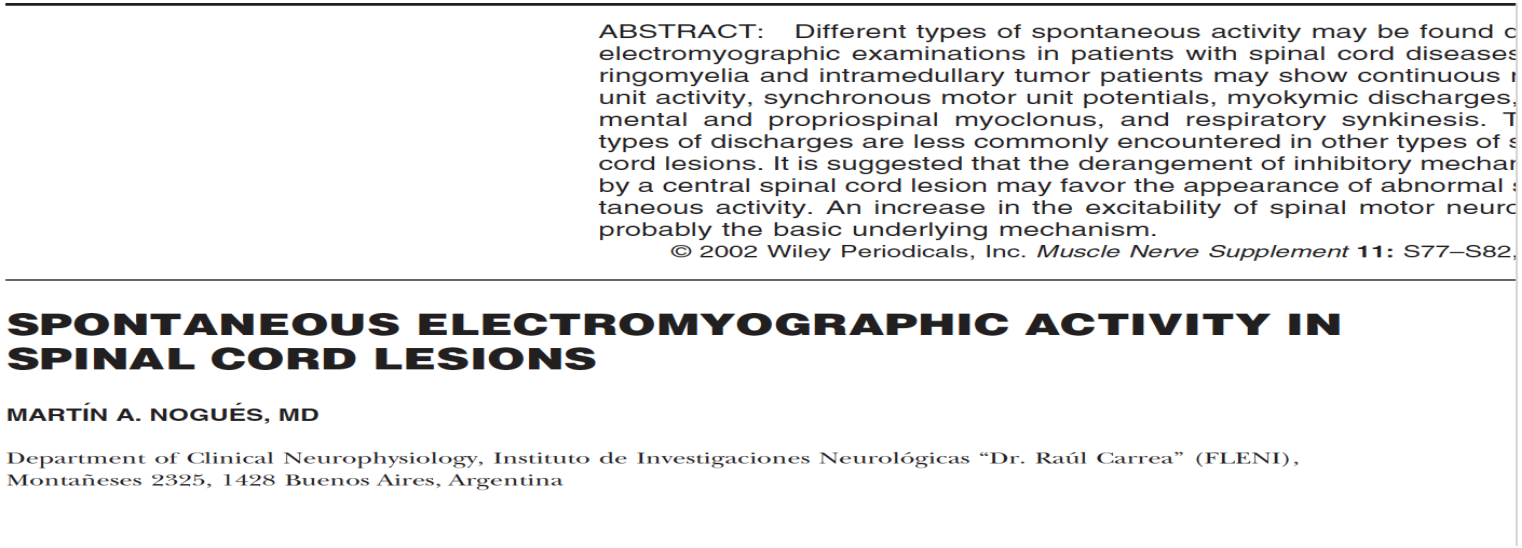



**FIGURE 6.** Left panel: mixed nerve silent period elicited by right (affected side) median nerve stimulation at the wrist during voluntary activation of the right APB muscle in a patient with dissociated sensory loss restricted to the right arm and upper trunk. Right panel: mixed nerve silent period evoked by stimulation of the left median nerve at the wrist and recording from the APB muscle in the same patient. The duration of the silent period on the right (normal) side is approximately double that on the affected side (surface recordings).



ABSTRACT: Different types of spontaneous activity may be found c
electromyographic examinations in patients with spinal cord diseases
ringomyelia and intramedullary tumor patients may show continuous r
unit activity, synchronous motor unit potentials, myokymic discharges,
mental and propriospinal myoclonus, and respiratory synkinesis. T
types of discharges are less commonly encountered in other types of s
cord lesions. It is suggested that the derangement of inhibitory mechar
by a central spinal cord lesion may favor the appearance of abnormal s
taneous activity. An increase in the excitability of spinal motor neuro
probably the basic underlying mechanism.
© 2002 Wiley Periodicals, Inc. *Muscle Nerve Supplement* **11:** S77–S82.

**SPONTANEOUS ELECTROMYOGRAPHIC ACTIVITY IN SPINAL CORD LESIONS**

**MARTÍN A. NOGUÉS, MD**

Department of Clinical Neurophysiology, Instituto de Investigaciones Neurológicas "Dr. Raúl Carrea" (FLENI), Montañeses 2325, 1428 Buenos Aires, Argentina

la componente spontanea anomala non è un dettaglio secondario, ma un marker del modo in cui la lesione midollare altera la fisiologia dei circuiti spinali

Sul piano pratico, questo significa che un quadro neurogeno con SNAP conservati, soprattutto se associato a fenomeni come CMUA, myokymia o sinchinesie respiratorie, deve orientare prioritariamente verso un interessamento intramidollare delle corna anteriori e dei circuiti segmentari, e non essere letto automaticamente come neuropatia periferica, radicolopatia o motoneuronopatia primaria

Naturalmente, la lettura deve restare sempre integrata con il contesto clinico e la RM, ma il valore dell'EMG sta proprio nel mostrare che la siringomielia, dal punto di vista funzionale, è molto più di una cavità compressiva o cicatriziale

## 23-05 H-Reflex Recovery Curve in Syringomyelia

C. Hoffman, H. García, A. Rivero, S. Estellés, M. Merello, M. Nogués. *FLENI, Buenos Aires*

The H-reflex recovery curve was assessed in 13 patients with a classic syringomyelic syndrome featuring dissociated sensory loss, lower motor neurone weakness in upper limbs and spastic paraparesis. Findings were compared with recordings from patients with spasticity due to other causes and from healthy control subjects. The purpose was to quantify motor neurone excitability and to correlate values with clinical and MRI findings. Throughout, diagnosis was confirmed by MRI.

The H-reflex was obtained by placing an active electrode over the belly of the flexor carpi radialis muscle. The median nerve was then stimulated at the elbow, using 13 double pulses automatically delivered by the EMG (E. and S. StaÅlberg, 1989).

The recovery curve was abnormal in 12 out of the 13 syringomyelia patients. Hyperexcitability was also commonly found on the non-affected side. Curves proved more anomalous in the syringomyelia than in the spastic group. The H-reflex was unobtainable from patients with proprioceptive sensory loss involving hands. The interruption of the reflex arc is most likely at the afferent limb. The H-reflex recovery curve may be useful to assess motor neurone excitability at different stages of syringomyelia.

Un discorso analogo vale per l'H-reflex recovery curve e, più in generale, per le metodiche che esplorano l'eccitabilità del circuito segmentario spinale. Il contributo di Hoffman ha un livello di evidenza limitato, ma il suo interesse è soprattutto fisiopatologico. Se la siringomielia altera non solo i tratti lunghi ma anche i meccanismi di controllo inibitorio segmentario, è plausibile che cambino anche i parametri che riflettono il recupero dell'eccitabilità del circuito riflesso. Queste tecniche non rappresentano necessariamente uno strumento di routine nella pratica quotidiana, ma hanno il merito di mostrare che il problema non è soltanto una lesione anatomica, bensì una riorganizzazione patologica della fisiologia spinale

Journal of
*Clinical Medicine*

MDPI

*Article*

# Neurophysiological Correlates in Patients with Syringomyelia and Chiari Malformation: The Cortico-Diaphragmatic Involvement

**Dario Cocito [1,2,*], Erdita Peci [2], Diego Garbossa [1,3] and Palma Ciaramitaro [1,3]**

[1] Department of Neuroscience "Rita Levi Montalcini", University of Torino, 10126 Torino, Italy
[2] ICS Maugeri, 10124 Torino, Italy
[3] CRESSC-Interregional Centre of Expertise for Syringomyelia and Chiari, Città della Salute e della Scienza University Hospital, 10126 Torino, Italy
* Correspondence: dario.cocito@icsmaugeri.it; Tel.: +39-3479736067

il danno funzionale può riguardare anche il controllo volontario della respirazione. Il dato che il Cz-MEP del diaframma risulti alterato in una quota di pazienti, e che il C5-MEP sia più spesso compromesso nei casi con siringomielia bulbo-cervicale associata a CM1, suggerisce che la lesione possa interferire non solo con i tratti lunghi, ma anche con il compartimento del secondo motoneurone correlato al nervo frenico o con le connessioni segmentarie cervicali. Il valore di questo lavoro non sta tanto nel proporre una nuova routine clinica, quanto nel ricordare che il coinvolgimento bulbo-cervicale può avere una traduzione funzionale più ampia di quanto non appaia alla sola osservazione clinica.

# Chirurgia

Qui è essenziale distinguere tra la decompressione della fossa posteriore per Chiari e la chirurgia diretta della siringa

Nelle **forme di CM1 non complesse**, l'IONM non ha ancora dimostrato un ruolo uniforme come pratica routinaria standard, e il beneficio più costantemente documentato riguarda soprattutto il posizionamento e alcuni casi selezionati, in particolare pediatrici o con anatomia della giunzione cranio-cervicale più complessa. Questo significa che nella decompressione della Chiari semplice il monitoraggio va considerato come strumento selettivo e contestuale

Journal of Clinical Medicine

MDPI

Article

# Intraoperative Neurophysiological Monitoring in Syringomyelia Surgery: A Multimodal Approach

M. Ángeles Sánchez Roldán [1,*], Dulce Moncho [1,2], Kimia Rahnama [1], Daniela Santa-Cruz [1], Elena Lainez [1], Daniel Baiget [1], Ivette Chocrón [3], Darío Gándara [4], Agustín Bescós [4], Juan Sahuquillo [2,4,5] and María A. Poca [2,4,*]

1 Department of Clinical Neurophysiology, Vall d'Hebron University Hospital, Passeig Vall d'Hebron 119-129, 08035 Barcelona, Spain; dulcemaria.moncho@vallhebron.cat (D.M.); kimia.rahnama@vallhebron.cat (K.R.); danielaisabel.santacruz@vallhebron.cat (D.S.-C.); daniel.baiget@vallhebron.cat (D.B.)
2 Neurotraumatology and Neurosurgery Research Unit, Vall d'Hebron Institut de Recerca (VHIR), Vall d'Hebron Barcelona Hospital Campus, Passeig Vall d'Hebron 119-129, 08035 Barcelona, Spain; sahuquillo@neurotrauma.net
3 Department of Anesthesiology, Vall d'Hebron University Hospital, Passeig Vall d'Hebron 119-129, 08035 Barcelona, Spain; ivette.chocron@vallhebron.cat
4 Department of Neurosurgery, Vall d'Hebron University Hospital, Passeig Vall d'Hebron 119-129, 08035 Barcelona, Spain; dario.gandara@vallhebron.cat (D.G.); agustin.bescos@vallhebron.cat (A.B.)
5 Department of Surgery, Universitat Autònoma de Barcelona, Bellaterra, 08193 Barcelona, Spain
* Correspondence: mariadelosangeles.sanchez@vallhebron.cat (M.Á.S.R.); pocama@neurotrauma.net (M.A.P.); Tel.: +34-93-274-62-41 (M.Á.S.R.); +34-93-489-35-14 (M.A.P.); Fax: +34-274-62-10 (M.Á.S.R.); +34-93-489-41-88 (M.A.P.)

la stabilità dei MEP si associa all'assenza di nuovi deficit, mentre la loro perdita persistente correla con peggioramento motorio postoperatorio

Nella chirurgia intramidollare diretta, il rischio più critico è spesso rappresentato dal danno al sistema corticospinale o alle strutture motorie funzionalmente collegate all'accesso chirurgico; per questo il MEP è la modalità che più direttamente intercetta l'endpoint clinico rilevante, cioè il danno motorio imminente

I SEP mantengono un ruolo importante ma più complementare, perché informano soprattutto sulle colonne posteriori;

il free-running EMG, invece, non misura la conduzione lungo un tratto, ma segnala una irritazione meccanica acuta a livello segmentario o radicolare

Ne deriva una gerarchia funzionale precisa: nella chirurgia diretta del syrinx, il MEP è il cardine del monitoraggio del rischio motorio, mentre SEP e free-running EMG aggiungono informazione complementare ma non equivalente

Journal of Clinical Medicine

MDPI

Article

# Intraoperative Neurophysiological Monitoring in Syringomyelia Surgery: A Multimodal Approach

M. Ángeles Sánchez Roldán [1,*], Dulce Moncho [1,2], Kimia Rahnama [1], Daniela Santa-Cruz [1], Elena Lainez [1], Daniel Baiget [1], Ivette Chocrón [3], Darío Gándara [4], Agustín Bescós [4], Juan Sahuquillo [2,4,5] and María A. Poca [2,4,*]

1 Department of Clinical Neurophysiology, Vall d'Hebron University Hospital, Passeig Vall d'Hebron 119-129, 08035 Barcelona, Spain; dulcemaria.moncho@vallhebron.cat (D.M.); kimia.rahnama@vallhebron.cat (K.R.); danielaisabel.santacruz@vallhebron.cat (D.S.-C.); daniel.baiget@vallhebron.cat (D.B.)
2 Neurotraumatology and Neurosurgery Research Unit, Vall d'Hebron Institut de Recerca (VHIR), Vall d'Hebron Barcelona Hospital Campus, Passeig Vall d'Hebron 119-129, 08035 Barcelona, Spain; sahuquillo@neurotrauma.net
3 Department of Anesthesiology, Vall d'Hebron University Hospital, Passeig Vall d'Hebron 119-129, 08035 Barcelona, Spain; ivette.chocron@vallhebron.cat
4 Department of Neurosurgery, Vall d'Hebron University Hospital, Passeig Vall d'Hebron 119-129, 08035 Barcelona, Spain; dario.gandara@vallhebron.cat (D.G.); agustin.bescos@vallhebron.cat (A.B.)
5 Department of Surgery, Universitat Autònoma de Barcelona, Bellaterra, 08193 Barcelona, Spain
* Correspondence: mariadelosangeles.sanchez@vallhebron.cat (M.Á.S.R.); pocama@neurotrauma.net (M.A.P.); Tel.: +34-93-274-62-41 (M.Á.S.R.); +34-93-489-35-14 (M.A.P.); Fax: +34-274-62-10 (M.Á.S.R.); +34-93-489-41-88 (M.A.P.)

Forse l'aspetto più interessante del lavoro di Sánchez Roldán è però il passaggio dal monitoring al mapping

Il dorsal column mapping e il root mapping non servono semplicemente a segnalare se si sta danneggiando qualcosa, ma aiutano a scegliere dove entrare e come orientare l'accesso in un modo funzionalmente più sicuro.

Questo cambia radicalmente la filosofia del monitoraggio intraoperatorio: non più sola sorveglianza passiva di un evento avverso, ma supporto attivo alla strategia chirurgica

È probabilmente qui che il ruolo della neurofisiologia nella siringomielia mostra la propria maturità più alta, perché il dato fisiologico non si limita a descrivere il danno, ma contribuisce direttamente a ridurlo

# Take home messages

- La siringomielia va considerata una patologia funzionale multicompartimentale.
- I BAEP e i SSEP appaiono particolarmente utili per documentare la sofferenza subclinica e definire meglio il follow-up dei pazienti incidentali o paucisintomatici;
- l'EMG fornisce i reperti più caratteristici del coinvolgimento segmentario e dell'iperccitabilità spinale;
- la TMS diaframmatica può aggiungere informazioni nei sottogruppi con interessamento bulbo-cervicale;
- l'IONM trova il suo razionale più forte quando l'intervento coinvolge direttamente il syrinx, mentre nella decompressione della Chiari non complessa mantiene un ruolo più selettivo, con utilità documentata soprattutto in casi scelti e per problematiche legate al posizionamento
- In definitiva, la neurofisiologia non sostituisce l'imaging né la decisione clinico-chirurgica, ma consente di restituire alla siringomielia la sua reale natura: quella di una mielopatia dinamica, nella quale anatomia e funzione devono essere lette in parallelo