



LE DISTONIE

Aggiornamento Clinico-terapeutico

Confronto Medico-Paziente



**Centro per le Tecnologie Innovative in
Neuroriabilitazione, Unità Complessa di
Riabilitazione Neurologica,**





OUTLINE

- **Aspetti fisiopatologici e basi neurofisiologiche della riabilitazione delle distonie**
- **Modalità proposte e dati sull'efficacia**
 - **Distonia cervicale**
 - **Distonia focale della mano**
 - **Altre distonie**
- **Prospettive future**

Definizione e classificazione delle distonie

Primarie
Eredo-degenerative
Secondarie

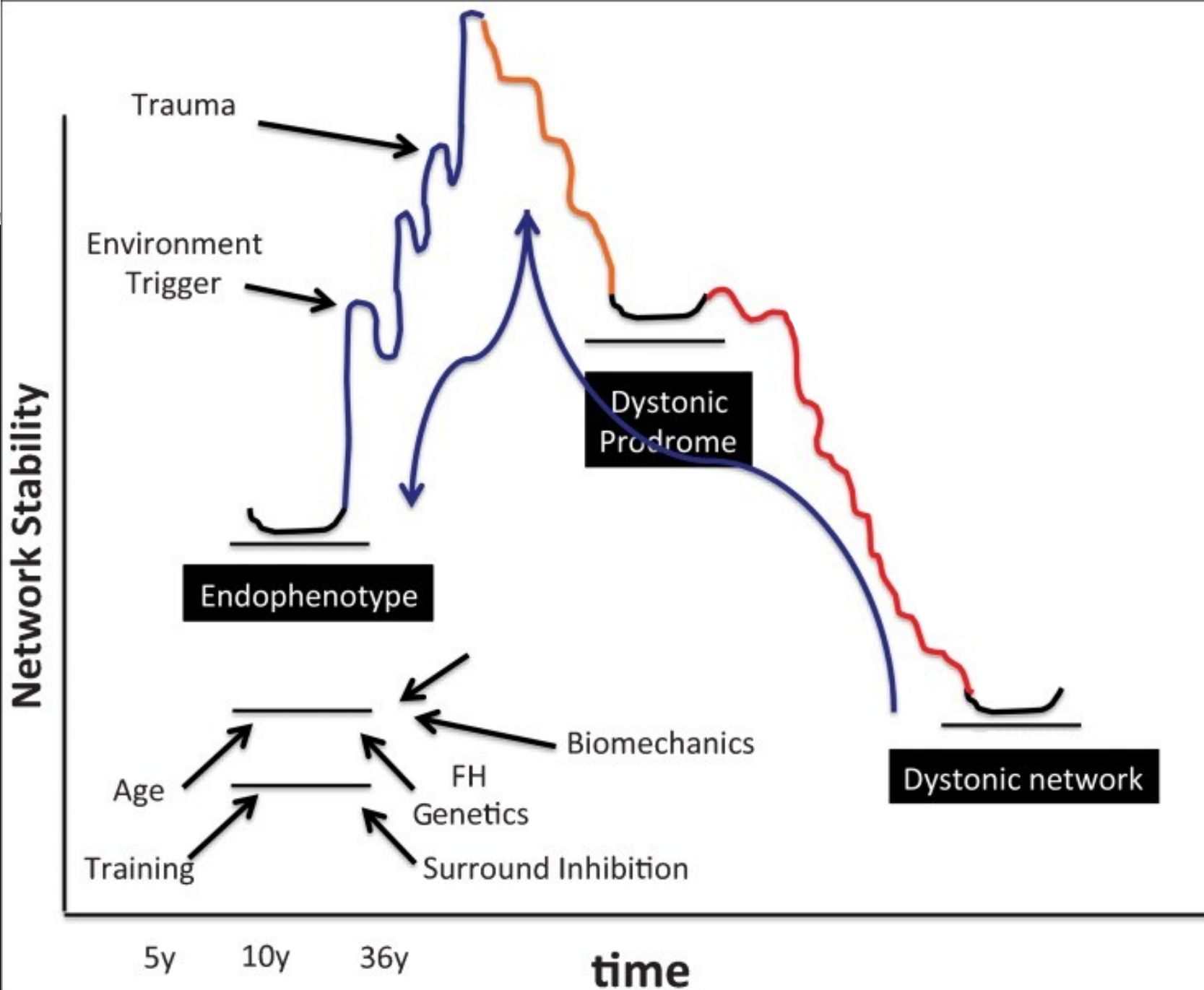
Focali
Segmentali
Generalizzate

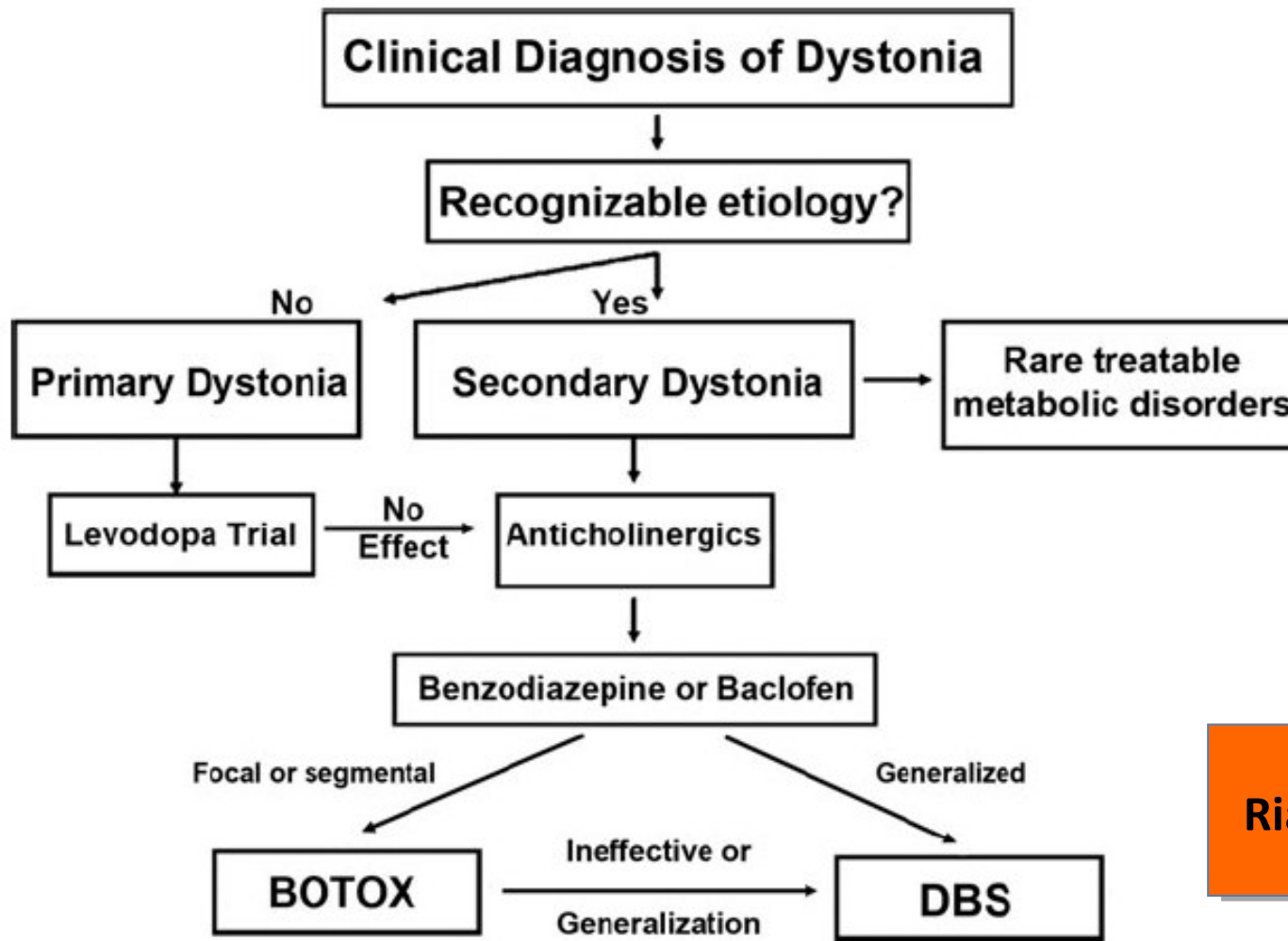




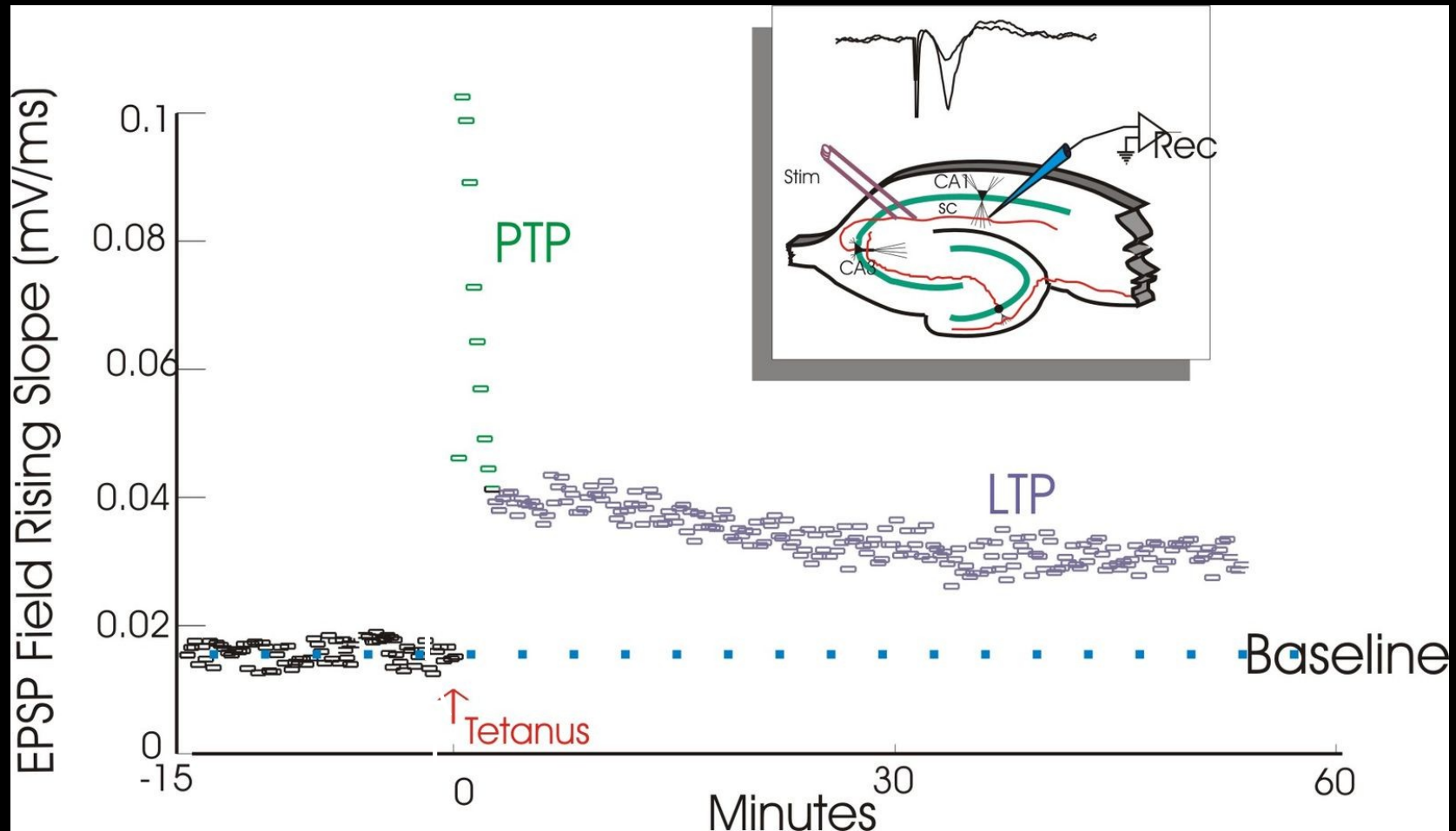
Aspetti fisiopatologici e basi neurofisiologiche	Modalità e dati di efficacia	Prospettive future
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SUBSTRATO FISIOPATOLOGICO





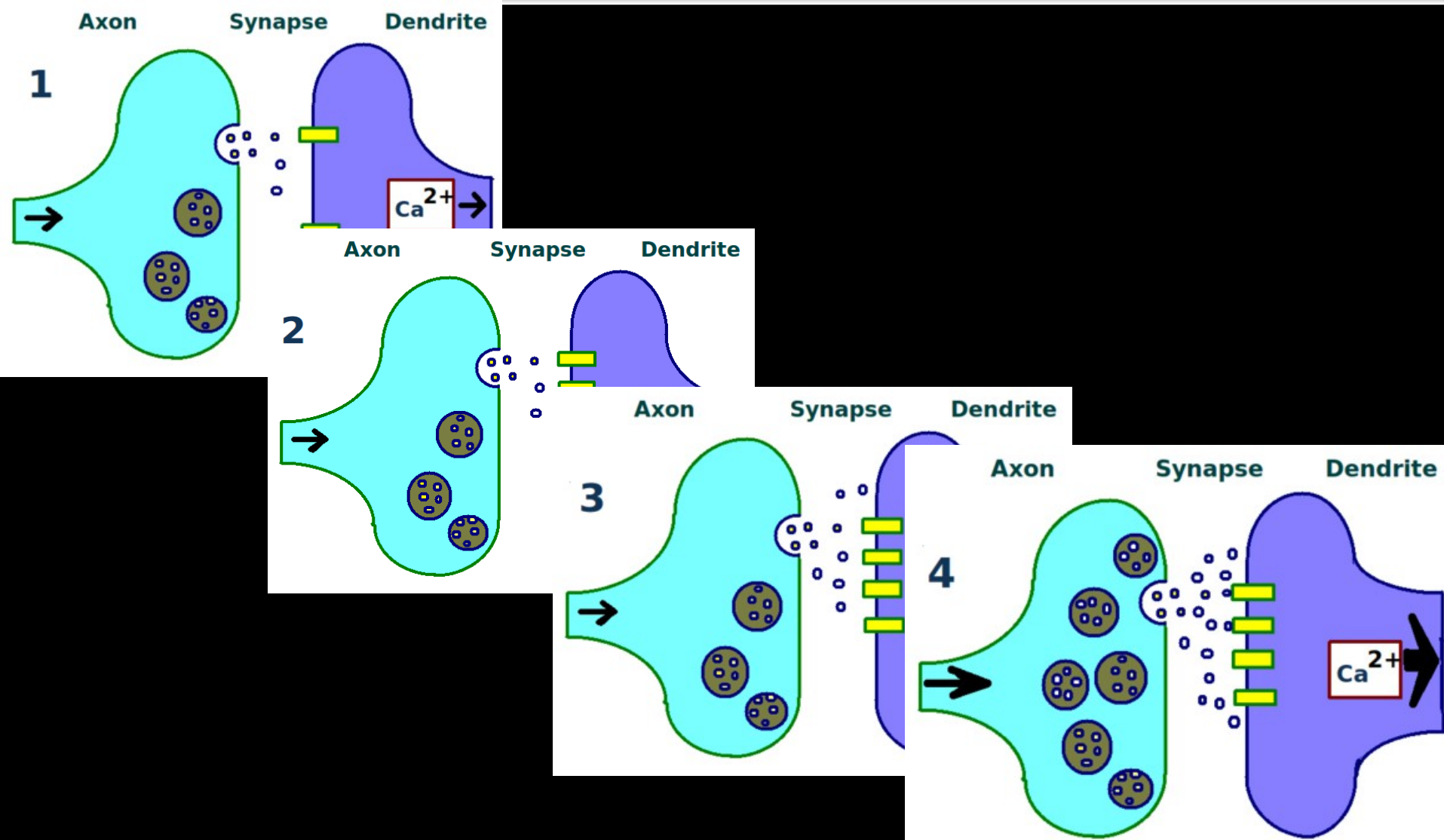
Riabilitazione



Aspetti fisiopatologici e
basi neurofisiologiche

Modalità e dati di
efficacia

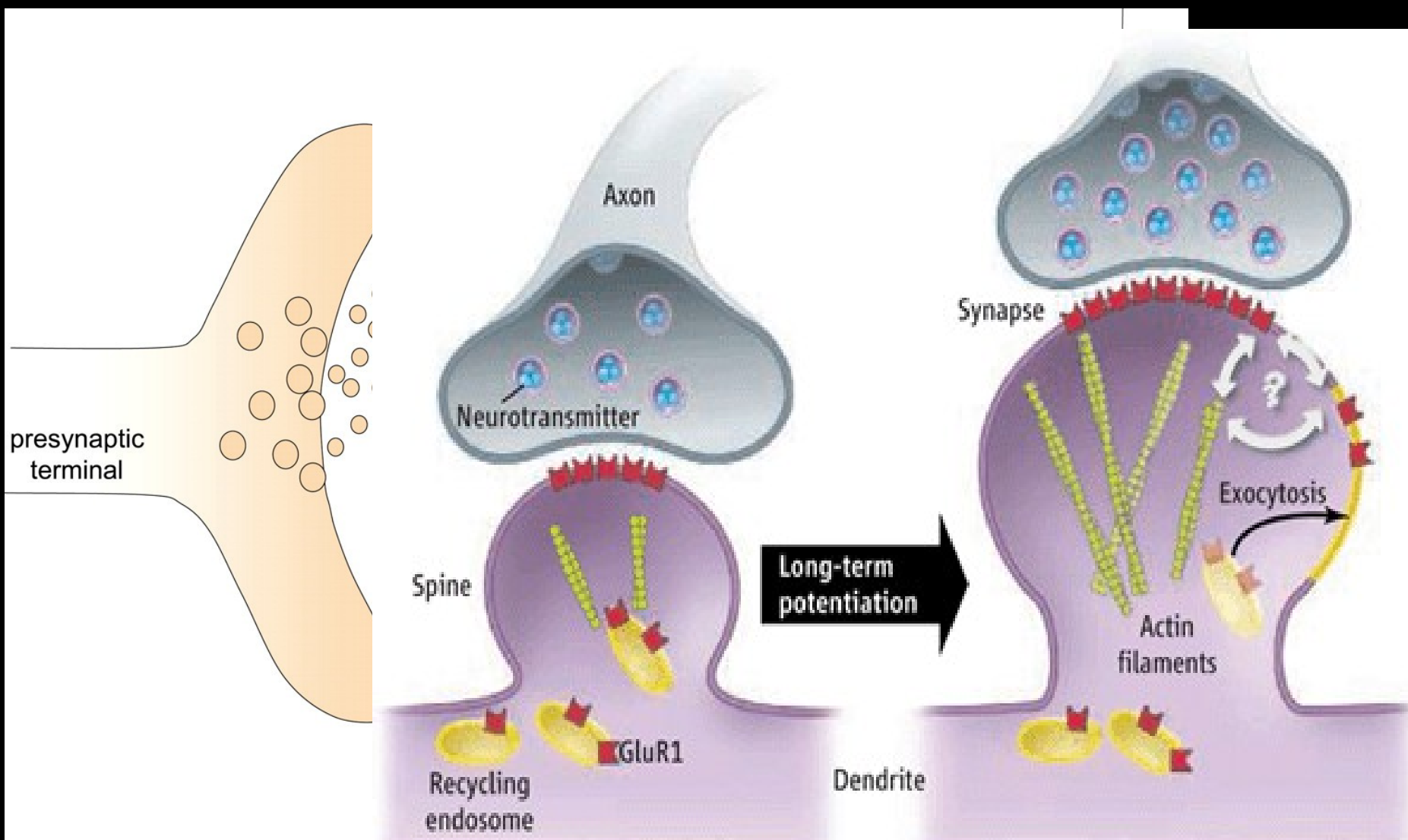
Prospettive
future

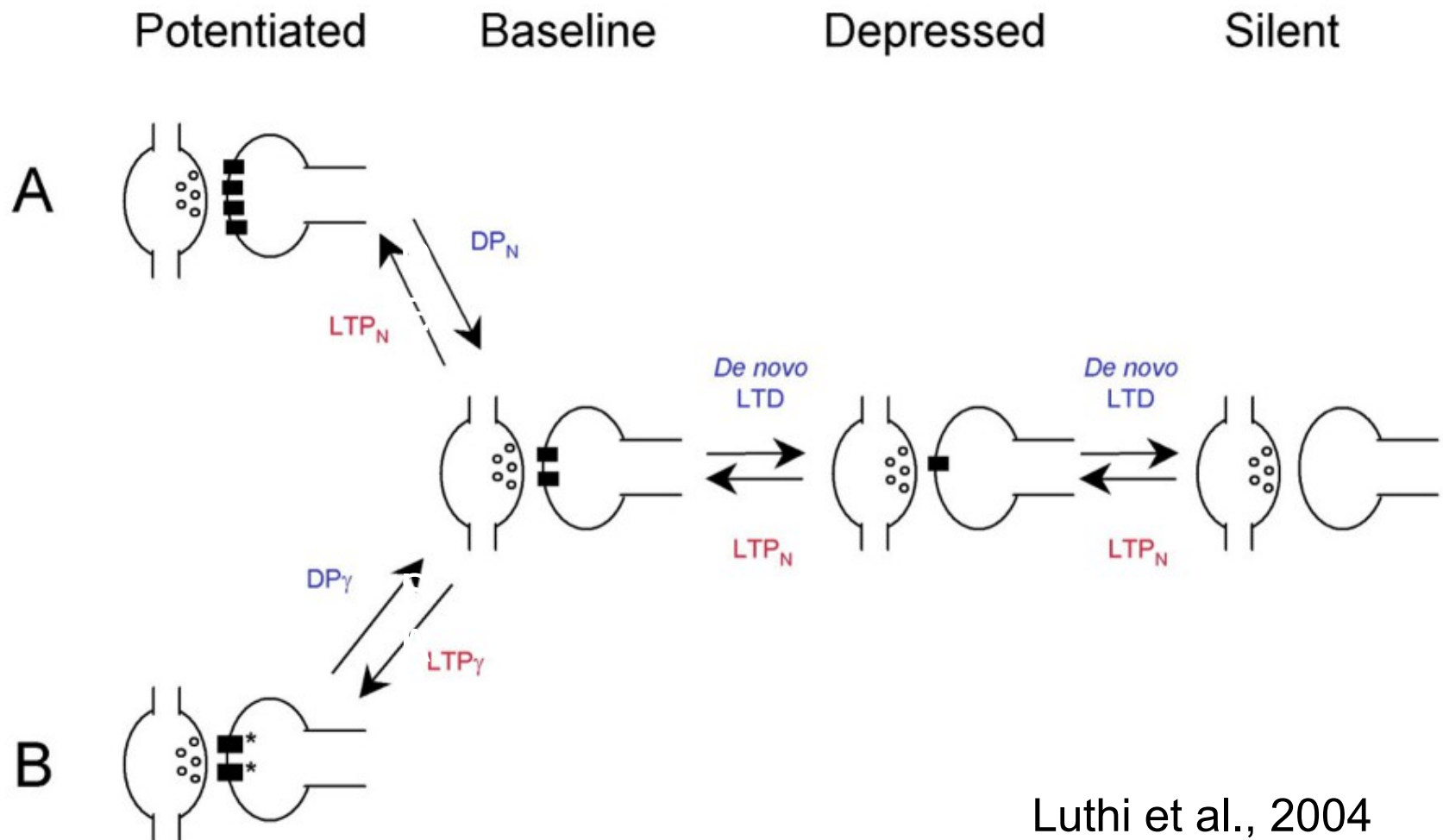


Aspetti fisiopatologici e
basi neurofisiologiche

Modalità e dati di
efficacia

Prospettive
future



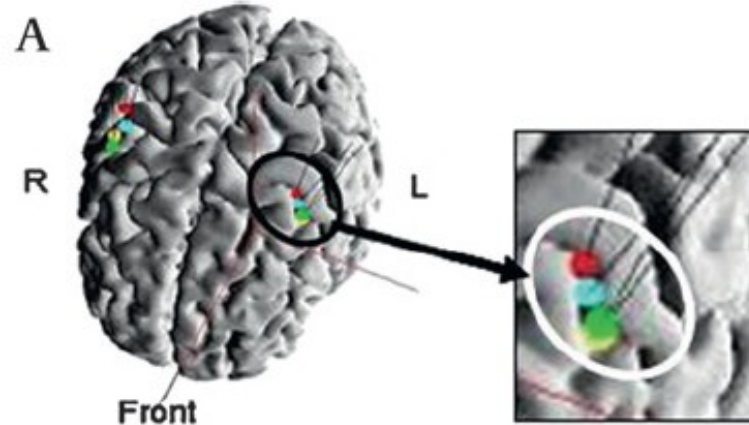




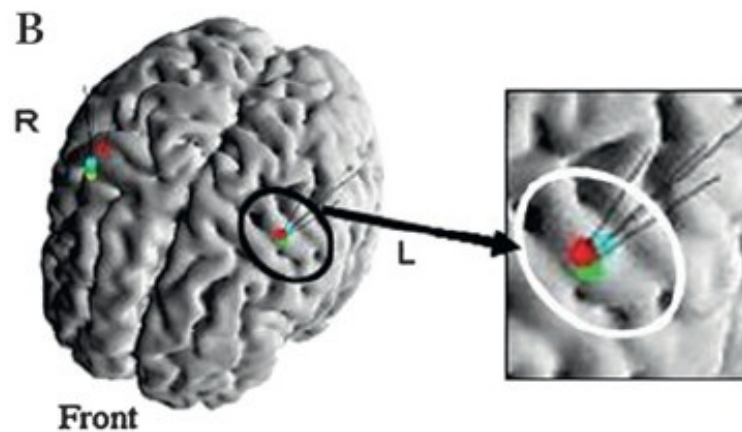
Aspetti fisiopatologici e basi neurofisiologiche	Modalità e dati di efficacia	Prospettive future
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Somatosensory cortical remodeling after rehabilitation and clinical benefit in writer's cramp

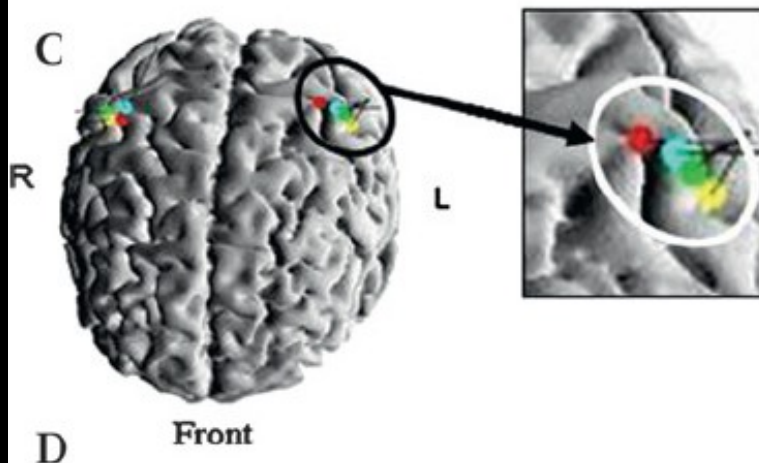
MEG and 3D-MRI



Healthy subject
normal
somatotopic order of
finger representation



Untreated WC
disorganised
somatotopic order of
finger representation



Rehabilitated WC
"super-normal"
somatotopic order of
finger representation

- Mignolo
- Medio
- Indice
- Pollice



Aspetti fisiopatologici e
basi neurofisiologiche

**Modalità e dati di
efficacia**

Prospettive
future

The effectiveness of physiotherapy for cervical dystonia: a systematic literature review

EMG biofeedback training

5 studi

2 soli di qualità buona

66 soggetti in tutto

Jahanshahi M, Sartory G, Marsden CD (1991) EMG biofeedback treatment of torticollis: a controlled outcome study. *Biofeedback Self Regul* 16:413–448

Tassorelli C, Mancini F, Balloni L et al (2006) Botulinum toxin and neuromotor rehabilitation: an integrated approach to idiopathic cervical dystonia. *Mov Disord* 21:2240–2243. doi:[10.1002/mds.21145](https://doi.org/10.1002/mds.21145)



Aspetti fisiopatologici e basi neurofisiologiche	Modalità e dati di efficacia	Prospettive future
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Aspetti fisiopatologici e
basi neurofisiologiche

**Modalità e dati di
efficacia**

Prospettive
future

Multimodal physiotherapy program

9 studi

1 di ottima qualità

3 di buona qualità

132 soggetti in tutto

Botulinum Toxin and Neuromotor Rehabilitation: An Integrated Approach to Idiopathic Cervical Dystonia

Tassorelli et al.,
Movement Disorders 2006



Popolazione

- 20 soggetti con DC

Tipo di studio

- Cross over randomizzato

Intervento/i

- Tossina botulinica
- Tossina botulinica +
fisioterapia

Durata

- 5 sedute alla settimana per 2
settimane

Indicatori di outcome

- TSUI
- TWSTRS



Aspetti fisiopatologici e
basi neurofisiologiche

**Modalità e dati di
efficacia**

Prospettive
future

TABLE 2. *Effect of botulinum toxin type A alone or in combination with physical therapy on different scales*

Parameter	BTX-0	BTX-PT
Tsui scale	-8.1 ± 3.2	-7.2 ± 2.8
TWSTRS	-9.1 ± 6.8	-10.2 ± 6.0
ADL	-5.3 ± 5.2	-9.8 ± 6.3^a
Total Pain score	-7.1 ± 3.2	-13.0 ± 3.2^b

^a $P < 0.05$.^b $P < 0.001$.

Queiroz et al., Funct Neurol 2012

N. 20

- Tossina botulinica
- Tossina botulinica +
fisioterapia

**Miglioramento in entrambi i
gruppi**

**Miglioramento più marcato del
dolore e della disabilità nel
gruppo tossina botulinica +
fisioterapia**

Boyce et al., Clin Rehab 2013

N. 20

- Rilassamento whole body
- Rilassamento + esercizi attivi

**L'inserimento degli esercizi
attivi causa solo una tendenza
verso un maggiore
miglioramento nella TWSTRS e
della depressione**

**El-baharawy et al.,
Egypt J Neurol Psychiatry Neurosurg 2009**

- **N. 40**
- **Stretching, esercizi attivi,
TENS e tossina botulinica +
fisioterapia**
- **TENS placebo + tossina
botulinica + fisioterapia**

**Migliore allineamento
posturale e migliore motilità
fine della mano nel gruppo con
stretching, es. attivi e TENS
vera**



Aspetti fisiopatologici e basi neurofisiologiche	Modalità e dati di efficacia	Prospettive future
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Aspetti fisiopatologici e
basi neurofisiologiche

**Modalità e dati di
efficacia**

Prospettive
future

SINTESI per Distonia cervicale

Review

Paramedical Treatment in Primary Dystonia: A Systematic Review

TABLE 1. *EBRO classification of study results according to the level of evidence*

-
- A1. Meta-analyses (systematic reviews), which include at least some, randomized clinical trials at quality level A2 that show consistent results between studies
 - A2. Randomized clinical trials of a good methodological quality (randomized double-blind controlled studies) with sufficient power and consistency
 - B. Randomized clinical trials of a moderate methodological quality or with insufficient power, or other nonrandomized, cohort or patient–control group study designs that involve intergroup comparisons
 - C. Patient series
 - D. Expert opinion
-

Reference, year of publication	No. (P,C)	Design	Intervention	Effect	EBRO
Ranawaya R, 1991 ¹⁸	20	Prospective case series, single blind	Writing device	75% improvement in dystonia, but in 9 patients not useful	C
Baur B, 2006 ¹⁹	23, 11	Prospective patient control study	Writing device	Reduced pen pressure, no effect on length or time	B
Koller WC, 1989 ²⁰	1	Case report	Writing device	Normal writing pattern without discomfort, longer duration of writing	D
Tas N, 2001 ²¹	5	Case series	Writing device	Improvement legibility hand writing, no discomfort	C
Lees AJ, 1989 ²²	1	Case report	Writing device	Longer comfortable writing	D
Espay AJ, 2005 ²³	9	Case series, single blind	Writing device	Improvement on writing	C
Deepak KK, 1999 ²⁷	10	Retrospective case series	Auditory EMG feedback	Decreased pain/discomfort objective/subjective	C
Berger HJ, 2007 ²⁶	5	Prospective case series, single blind	Auditory and visual EMG feedback and SPECT	Improvement of writing with relaxed limb; D2-binding restoration on SPECT	C
O'Neill MA, 1997 ²⁵	1	Case report	Auditory and visual EMG-feedback	Subjective improvement in writing comfort, reduction EMG-activity	D
Cottraux JA, 1983 ²⁸	13	Case series	9× Behavior therapy package + EMG feedback 4× Auditory EMG feedback	4 dropouts; 9 improvements in writing performance	C
Zeuner KE, 2002 ³³	10, 10	Prospective case series	Sensory discrimination therapy	Improved sensory perception correlated positively with improvement on FDS	B
Zeuner KE, 2003 ³⁴	3	Follow-up 2002	Sensory discrimination therapy	More and persistent improvement in 2 patients after continuing therapy	C
Tinazzi M, 2005 ³⁵	10	Prospective case series, crossover, randomized double blind	TENS versus placebo	Improvement in writing time and subjective assessment of writing, not in DMS	B
Toglia JU, 1985 ⁸¹	1	Case report	TENS	Disappearance of tremor during TENS, regression after 15 minutes	D
Schenk T, 2004 ²⁴	29, 21	Prospective case control study	Individual hand writing training	Significant improvement in writing performance	B
Zeuner KE, 2005 ³⁶	10	Prospective case series	Motor training	15–80% improvement subjectively in 6/10, significant improvement on FDS	C
Berg D, 1999 ¹⁷	26	Retrospective case series, crossover design	Occupational program + BoNT-A versus occupational program	15 discontinued Rx; 58% response to BoNT-A in various degrees; 33% marked/moderate improvement with OT	B
Pohl C, 2002 ²⁹	10, 10	Prospective patient control study, single blind	Cooling and warming arm	Mild improvement-no change subjective assessment, better writing performance	B

Sensory Training for Patients with Focal Hand Dystonia

Zeuner et al., 2002



Popolazione

- 10 soggetti con crampo dello scrivano

Tipo di studio

- Controllato vs soggetti sani

Intervento

- Allenamento mediante uso di alfabeto Braille

Durata

- 8 settimane, sedute quotidiane della durata di 30-60 minuti

Indicatori di outcome

- Percezione sensoriale
- Fahn dystonia scale
- Velocità di scrittura di paragrafo
- Scala visiva e verbale di miglioramento soggettivo

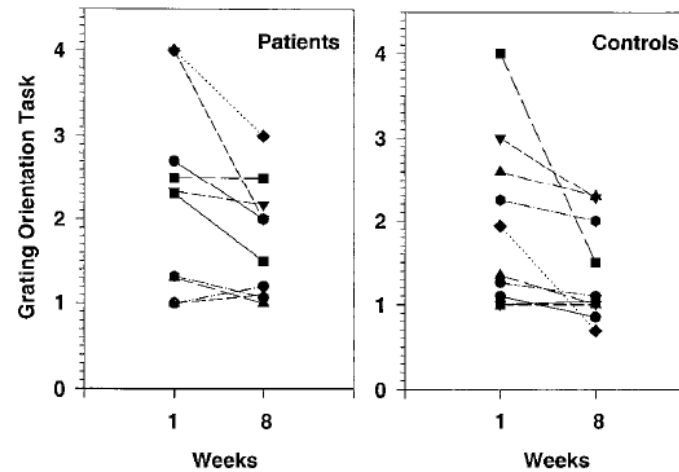


Fig 1. Thresholds, in millimeters, in the grating orientation task at baseline and after week 8 for 10 patients (left) and 10 normal volunteers (right). Two patients and 1 normal volunteer could not recognize the grating with 3mm at baseline; they were given an arbitrary number of 4mm.

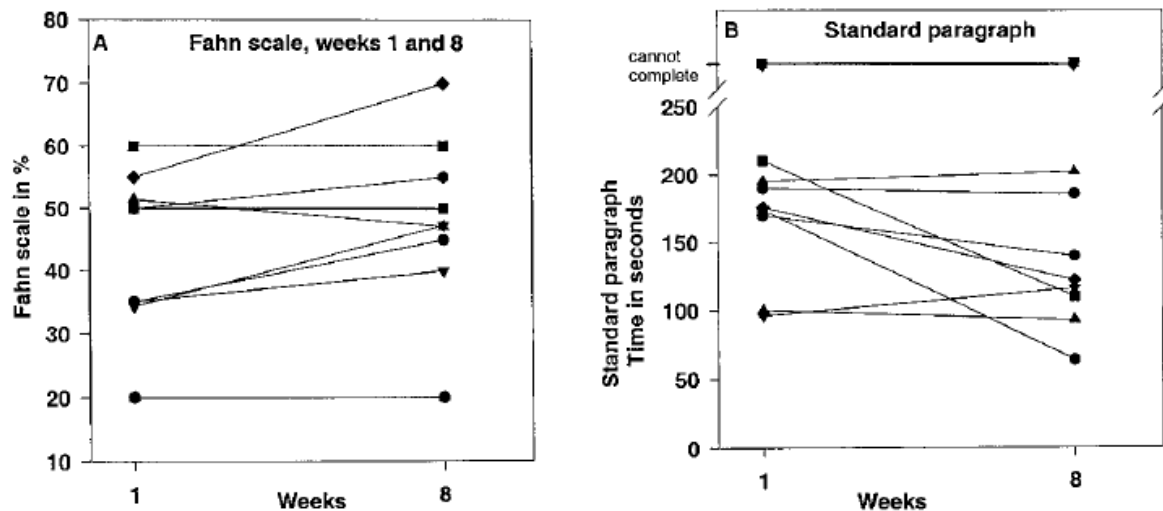


Fig 2. (A) Fahn dystonia scale scores for 10 dystonia patients at baseline and at week 8. A score of 100% meant lack of awareness of any disability. (B) Time taken to write a standard paragraph, recorded in seconds, for 10 patients at baseline and at week 8. Patients 4 and 8 could not complete the writing. They were given an arbitrary, unchanged number of 300 seconds.

Motor Training as Treatment in Focal Hand Dystonia[†]

Zeuner et al., 2005

Popolazione

- 10 soggetti con crampo dello scrivano

Tipo di studio

- Controllato vs soggetti sani

Intervento

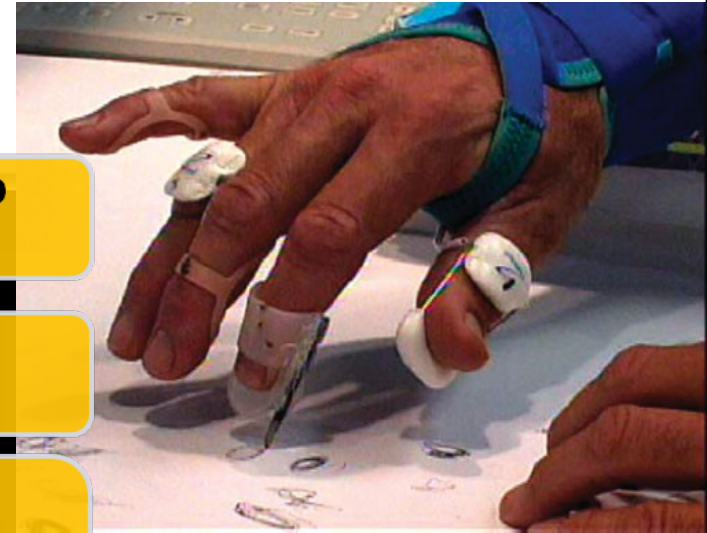
- Allenamento mediante training motorio (cerchi, ll, mm)

Durata

- 4 settimane, sedute quotidiane della durata di 20-50 minuti

Indicatori di outcome

- Analisi cinematica della scrittura (pressione e frequenza)
- Velocità di scrittura
- Fahn dystonia scale



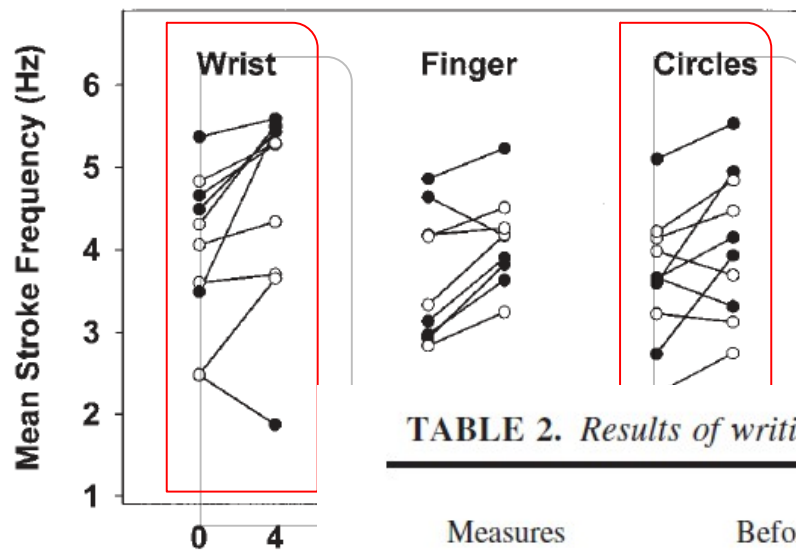


TABLE 2. Results of writing parameters before and after 4 weeks of training

Measures	Before motor training	After 4 weeks of motor training	<i>P</i>
Standard sentence			
Time (sec)	10885.30 ± 3862.75	9523.60 ± 3120.27	0.084
Pressure (n)	2.71 ± 0.89	2.95 ± 0.61	0.322
Frequency (Hz)	3.05 ± 0.76	3.16 ± 0.80	0.496
Lower case II			
Pressure (n)	2.59 ± 0.94	2.76 ± 0.90	0.432
Frequency (Hz)	2.67 ± 0.78	2.83 ± 0.65	0.557
Wrist movements			
Pressure (n)	2.25 ± 0.75	2.77 ± 0.78	0.232
Frequency (Hz)	3.98 ± 0.97	4.61 ± 1.22	0.020
Finger movements			
Pressure (n)	2.85 ± 1.00	3.10 ± 0.89	0.203
Frequency (Hz)	3.46 ± 1.00	3.81 ± 1.08	0.084
Circles			
Pressure (n)	2.65 ± 1.20	3.16 ± 0.83	0.232
Frequency (Hz)	3.65 ± 0.80	4.07 ± 0.89	0.037

Values are expressed as mean ± SD. All comparisons were performed using the Wilcoxon signed rank test. *P* values in boldface type are statistically significant.

Motor Re-Training Does Not Need to Be Task Specific to Improve Writer's Cramp

Zeuner et al., 2008

Popolazione

- 26 soggetti con crampo dello scrivano

Tipo di studio

- Controllato vs soggetti sani (21)

Intervento/i

- Allenamento mediante training motorio task-specific (scrivere lettere, disegnare cerchi)
- Allenamento mediante training motorio non task specific (uso di pongo)

Durata

- 8 settimane, sedute quotidiane della durata di 20-35 minuti

Indicatori di outcome

- Writer's cramp rating scale
- Arm dystonia disability scale
- Kinematic analysis

A



B



C



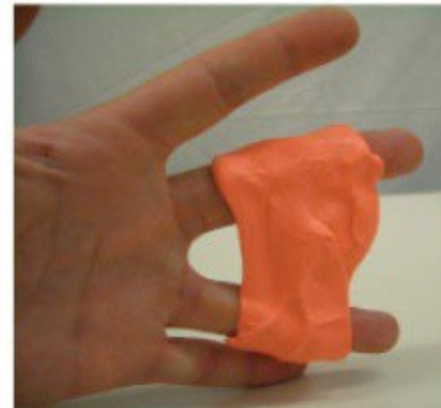
D

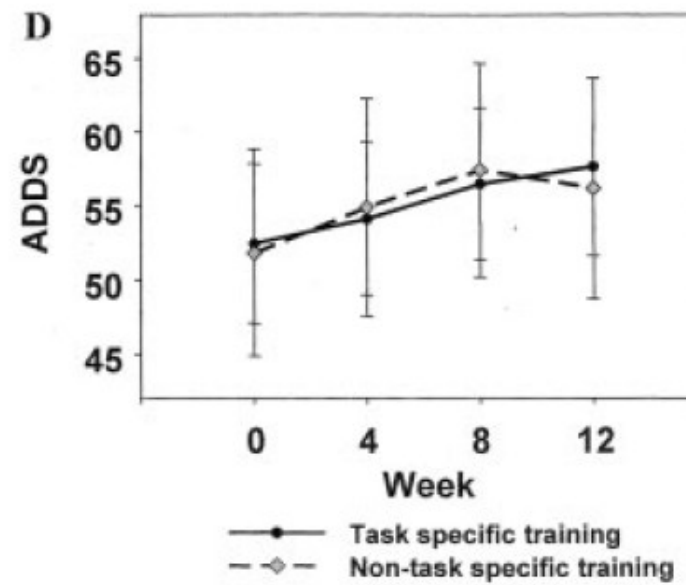
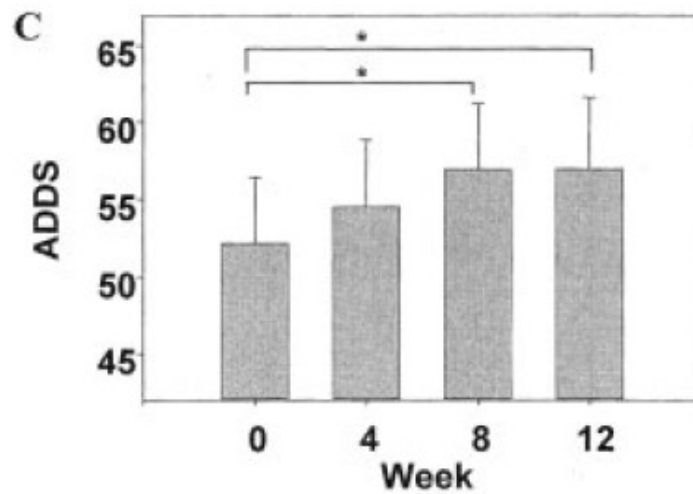
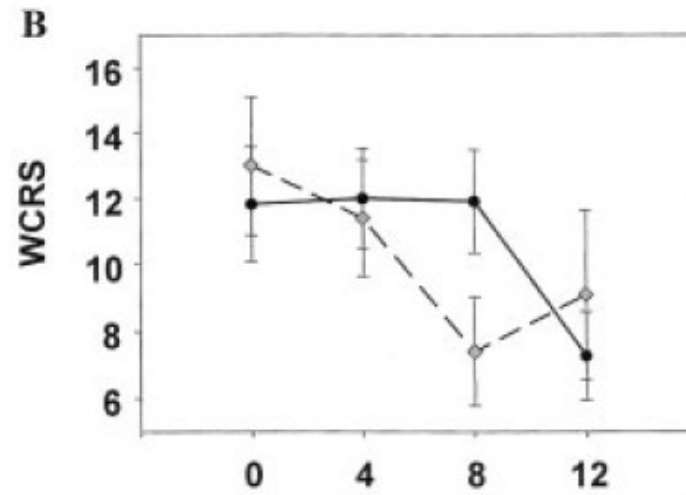
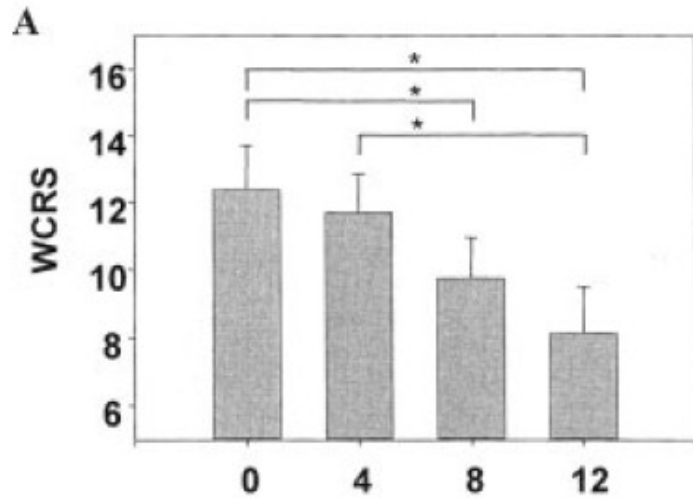


E



F





Reference, year of publication	No. (P,C)	Design	Intervention	Effect	EBRC
Candia V, 2002 ⁴³	11	Prospective case series	Sensory motor retuning therapy	Pianist and guitarist: significant improvement in performance. Wind players: no substantially improvement.	C
Candia V, 1999 ⁴⁴	5	Case series	Sensory motor retuning therapy	1 noncompliant regression, 3 improvement to normal, 1 no effect.	C
Byl NN, 2000 ⁴⁶	12	Prospective case series	Sensory discrimination rehabilitation program	Significant gain in pain, posture, balance, neural tension, strength, range of motion, sensory function, and motor accuracy	C
Byl NN, 2003 ⁴⁰	3, 30	Prospective case series. Normative controls	Individual rehabilitation program with repetitive sensorimotor training activities	Improved average target-specific performance, fine motor skills, sensory discrimination, musculoskeletal skills, and independence.	C
Sakai N, 2006 ⁴⁷	20	Prospective case series	Retraining: Slow down exercises	Improvement in ADDS and TCS	C
Ackermann BJ, 2005 ⁸²	1	Case report	Sensorimotor retraining using pseudocello	Improvement Candia scale, improvement in TCS	D
De Lisle R, 2006 ³⁷	3	Case series	Rehabilitation	Improvement in DER and PER	C
Tubiana R, 2003 ³⁹	145	Retrospective case series	Rehabilitation program	35 failures to complete, 25 no improvement, 85 mild improvement of which 39 return to performing	C
Jabusch HC, 2005 ³⁸	144	Retrospective case series	Various treatment strategies	Improvement in 63% for ergonomic changes; 50% for pedagogical training; 56% nonspecific technical exercises	C
Schuele S, 2004 ⁸³	18	Retrospective case series	Various treatment strategies	PT/retraining/biofeedback: none to moderate improvement; BoNT-A: 2 partial improvement; splint mild improvement	C
Levine WR, 1983 ⁸⁴	1	Case report	Thermometric EMG feedback	Subjective improvement persistent for 4 years	D
Priori A, 2001 ⁴¹	8	Prospective case series	Immobilizing therapy	Clumsiness hand, decubitus, weakness→ regain normal status, improvement dystonic posture, variable persistence of outcome	C
Pesenti A, 2004 ⁴²	19	Prospective case series	Immobilizing therapy	Clumsiness hand, decubitus, weakness→ regain normal status, highly variable improvement dystonic posture and, duration of effect	C

ADDS, arm dystonia disability scale; TCS, Tubiana and Chamagne scale; DER, dystonia evaluation rate; PER, pianists evaluation rate.

Prolonged Neuromuscular Rehabilitation for Musician's Focal Dystonia

Tubiana, 2003



Popolazione

- 145 soggetti con crampo del musicista

Tipo di studio

- Aperto

Intervento

- Ristrutturazione dell'immagine corporea
- Indipendenza dei movimenti dell'arto
- Rieducazione del movimento e della postura
- Rieducazione all'uso dello strumento

Durata

- Non specificato, ma fino a 2 anni

Indicatori di outcome

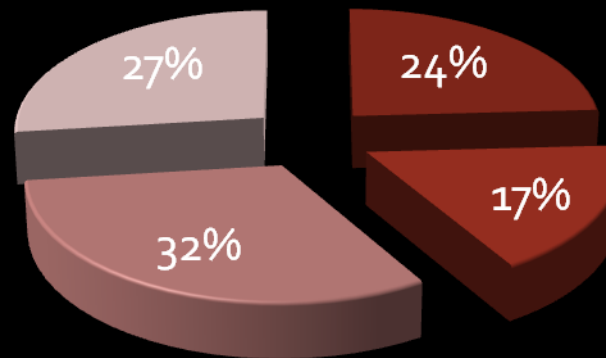
- Miglioramento soggettivo

Aspetti fisiopatologici e
basi neurofisiologiche

**Modalità e dati di
efficacia**

Prospettive
future

145 soggetti



- Drop out
- Nessuna efficacia
- Efficacia parziale
- Efficacia completa

Popolazione

- 5 soggetti con crampo del musicista

Tipo di studio

- Aperto

Intervento

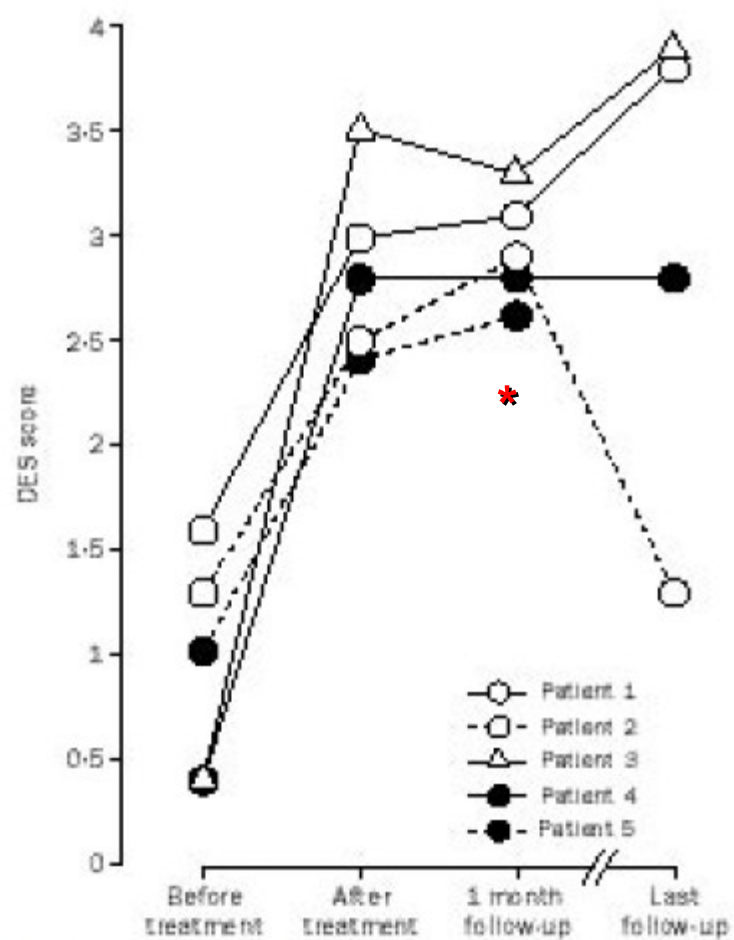
- Immobilizzazione di una o due dita adiacenti a quello distonico associata a training motorio

Durata

- 4 settimane

**Indicatori di
outcome**

- Dystonia evaluation scale



DES scores before and after treatment in five patients

Blefarospasmo e distonia oromandibolare

Reference, year of publication	No. (P,C)	Design	Intervention	Effect	EBRO
Rowan G, 1981 ⁴⁸	1	Case report	Skin temperature biofeedback and auditory EMG feedback	Absence spasms for 30 minutes, reduction EMG-activity	D
Bertolotti G, 2005 ⁴⁹	2	Case report	Auditory EMG feedback with relaxation exercises	Decreased EMG level of spasm frequency to asymptomatic	D
Peck DF, 1977 ⁵⁰	1	Case report	Auditory EMG feedback	Reduction in EMG activity and spasm frequency; control of spasms after 4 months	D
Roxanas MR, 1978 ⁵¹	1	Case report	Auditory and visual EMG feedback	Improvement to normal functioning, reduction in EMG activity; mild regression after 6 months	D
Brantley PJ, 1985 ⁵²	1	Case report	5.5 weeks Artane followed by auditory EMG-feedback	Reduction in EMG activity; reduction in eye blinking in EMG-BF and Artane	D
Stephenson NL, 1976 ⁵³	1	Case report	Relaxation therapy and EMG feedback	Improvement, after 6 months of no treatment asymptomatic	D
Bird BL, 1978 ⁵⁴	1	Case report	Auditory and visual EMG feedback	Marked improvement in voluntary spasm control	D
Sharpe R, 1974 ⁵⁵	1	Case report	Relaxation therapy	Reduction in spasm frequency without relapse after 9 months	D
Reinders M, 1989 ⁵⁶	1	Case report	Behavior therapy	78% reduction in eye blinking, relaxation facial musculature	D
Penaroccha M, 2001 ⁵⁷	1	Case report	Mandibular overdenture and endosteal implants	Stable occlusion and improved chewing	D
Penaroccha M, 2001 ⁵⁸	1	Case report	Mandibular overdenture and endosteal implants	Stable occlusion and improved chewing	D



Aspetti fisiopatologici e
basi neurofisiologiche

**Modalità e dati di
efficacia**

Prospettive
future

SINTESI PER DISTONIE FOCALI DELLA MANO

Intervento/i

- Allungamento miofasciale passivo e attivo
- Correzione e controllo posturale
- Potenziamiento dei muscoli ad azione antagonista della postura
- Biofeedback

Durata

- A cicli (prolungati nel crampo del musicista), da ripetere



Aspetti fisiopatologici e basi neurofisiologiche	Modalità e dati di efficacia	Prospettive future
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**Studi clinici controllati, anche a
lungo termine**

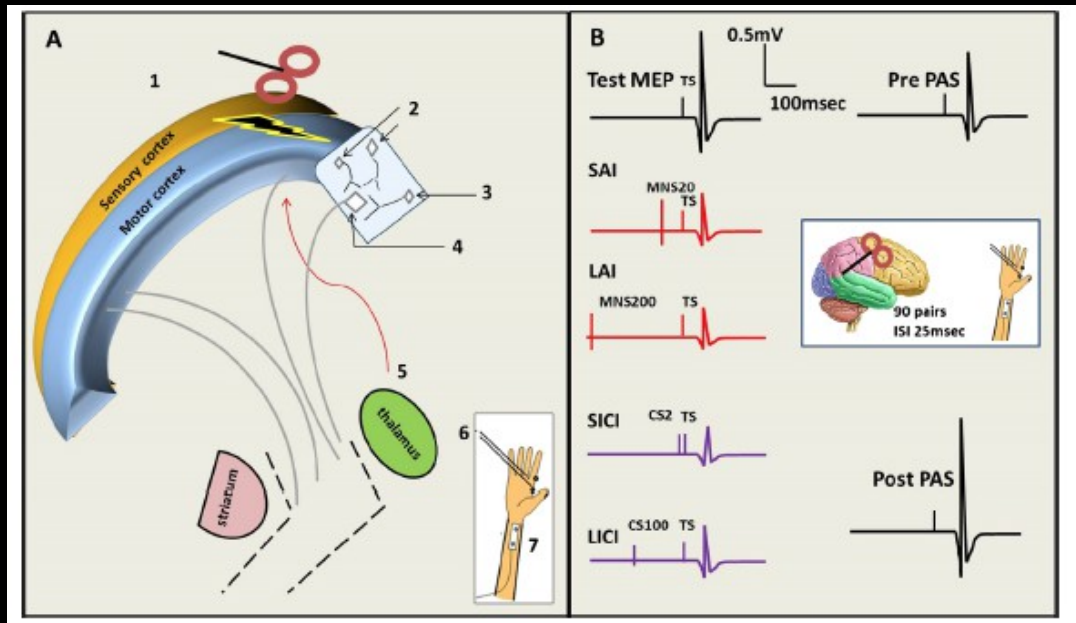
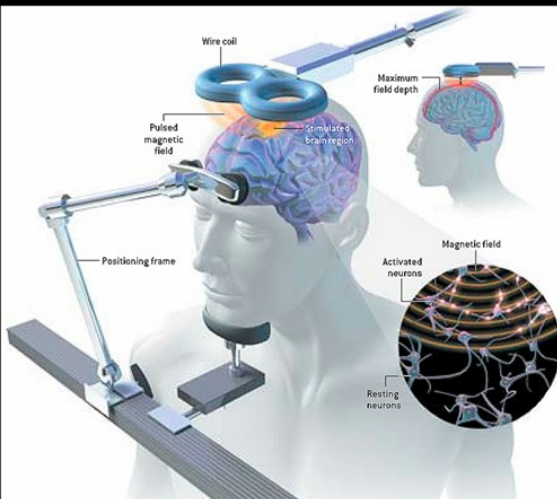
Studi clinici N=1 ben condotti

Aspetti fisiopatologici e
basi neurofisiologiche

Modalità e dati di
efficacia

Prospettive
future

rTMS



Aspetti fisiopatologici e basi neurofisiologiche

Modalità e dati di efficacia

Prospettive future

TABLE 5. Repetitive TMS Studies in Dystonia Patients with Clinical Outcome Measures

Study	Design	Extrinsic Factors			Intrinsic Factors		
		TMS Settings	Sessions and Duration	Pulses, no.	Subjects	Dystonia type	Outcome Measures
Siebner et al. ¹¹⁷ (1999)	Within-subject, real vs control rTMS	Figure-8 coil; M1 1 Hz at 90% MT	1 session in 30 min	1800	16 patients; 11 controls	Writer's cramp	Handwriting measures (+); also excitability
Lefaucheur et al. ¹²⁶ (2004)	Open-label case-reports	Figure-8 coil; PMd 1 Hz at 90% MT	5 sessions in 5 d	6000	3 patients, severe painful, dystonia	Generalized, secondary dystonia	Reduced pain dramatically; slight changes in dystonia rating scales
Murase et al. ¹²⁵ (2005)	Within-subject, real vs control rTMS	Figure-8 coil M1, PMd, SMA 0.2 Hz at 85% MT	1 session in 21 min	250/site	9 patients	Writer's cramp	Handwriting measures (+) over PMd only; also excitability
Allam et al. ¹²⁷ (2007)	Open-label, case report	Figure-8 coil; PMd 1 Hz at 90% MT	5 sessions in 5 d	6000	1 patient	Cervical dystonia and writer's cramp	Cervical dystonia (+); writer's cramp (0)

**Aspetti fisiopatologici e
basi neurofisiologiche**

**Modalità e dati di
efficacia**

**Prospettive
future**

Lasting effects of repeated
rTMS application in focal
hand dystonia. Borich et al.,
2009

Popolazione

- 5 soggetti con
distonia focale
della mano e 9
controlli sani

**Tipo di
studio**

- Controllato

Intervento

- rTMS (1Hz)

Durata

- 5 sedute (1 sola
nei controlli)

**Indicatori di
outcome**

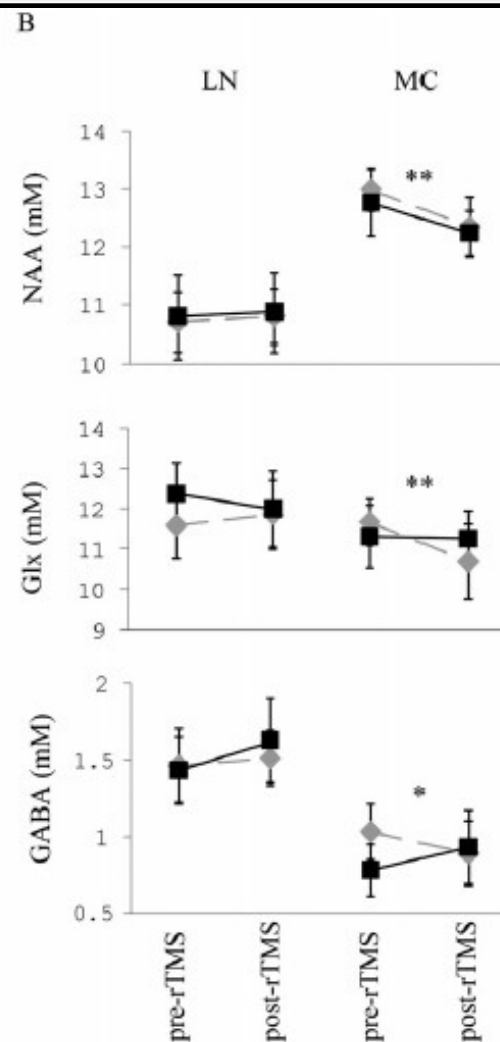
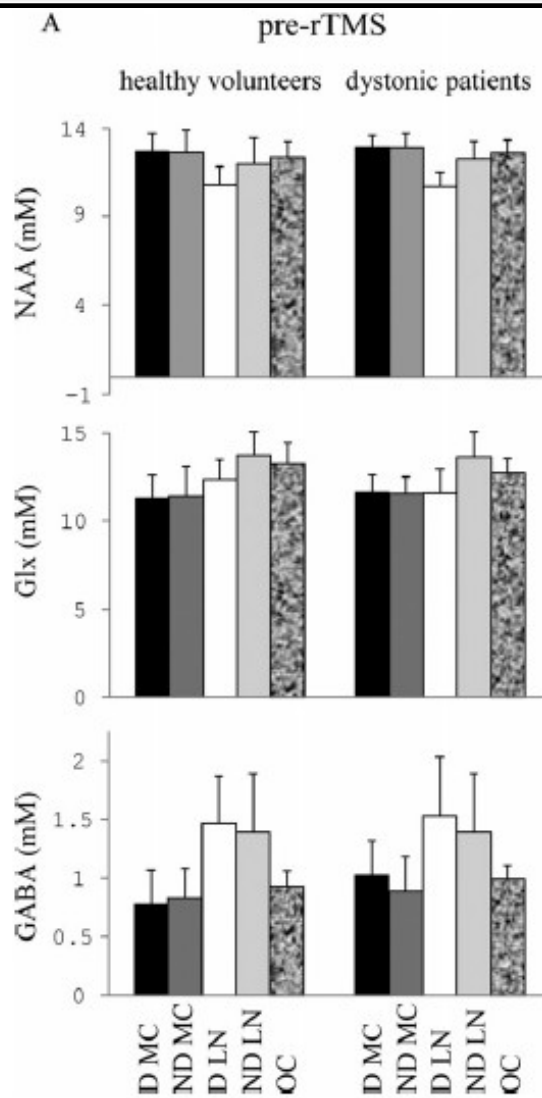
- Periodo silente
corticale
- Parametri di
scrittura

Aspetti fisiopatologici e basi neurofisiologiche

Modalità e dati di efficacia

Prospettive future

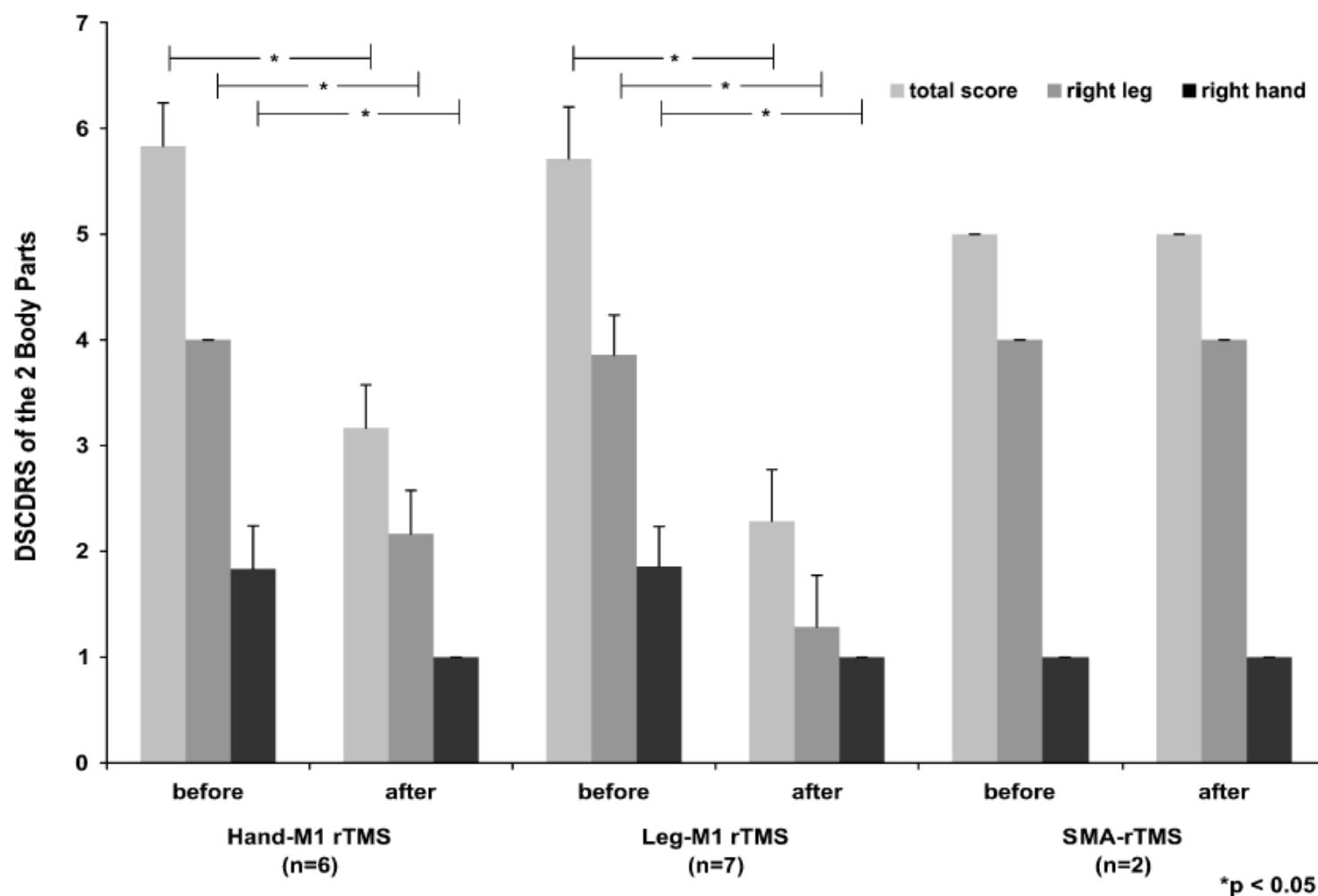
rTMS sulla corteccia motoria



Magnetic Resonance Spectroscopy

Marjans
Mov Dis

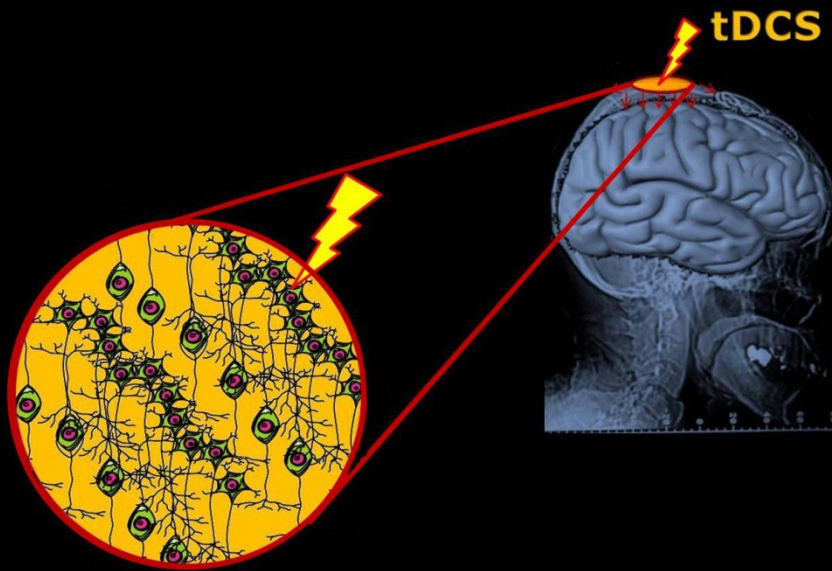
Kodama M, Kasahara T, Hyodo M, Aono K, Sugaya M, Koyama Y, Hanayama K, Masakado Y: Effect of low-frequency repetitive transcranial magnetic stimulation combined with physical therapy on L-dopa-induced painful off-period dystonia in parkinson's disease. *Am J Phys Med Rehabil* 2011;90:150–155.



**Aspetti fisiopatologici e
basi neurofisiologiche**

**Modalità e dati di
efficacia**

**Prospettive
future**



(Priori, 1998 e 2003)

Transcranial alternating current stimulation reduces symptoms in intractable idiopathic cervical dystonia: a case study.

Angelakis et al. Neurosci Lett. 2013

Blinded, placebo-controlled case study, transcranial direct current stimulation (tDCS) and transcranial alternating current stimulation (tACS) has been applied to a 54-year old patient with intractable ICD.

Results showed that 15 Hz tACS had both immediate and cumulative effects in dystonic symptom reduction, with a 54% reduction in the TWSTRS total score, and a 75% in the TWSTRS Pain Scale.

These effects were persistent at 30-days follow-up.

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RIABILITAZIONE NELLA DISTONIA

Farmaci

**Neurochirurgia
funzionale**

**Tossina
botulinica**