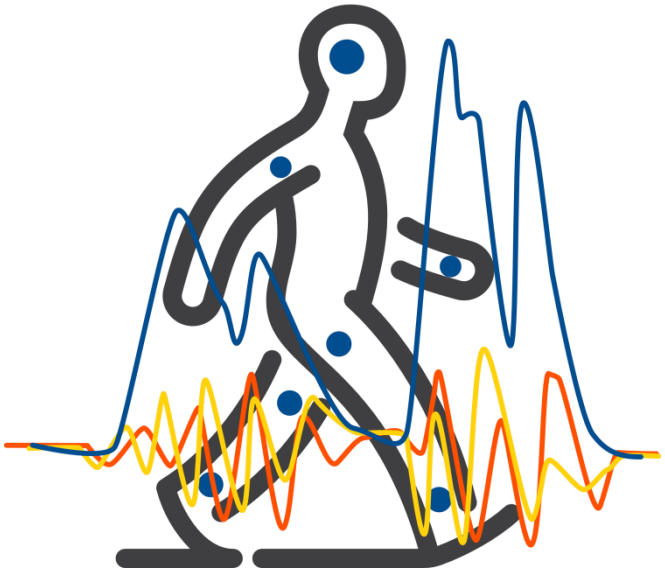
European Reference Network for rare or low prevalence complex diseases
Network Neurological Diseases (ERN-RND)

ean european academy of neurology

European Reference Network for rare or low prevalence complex diseases
Network Neuromuscular Diseases (ERN EURO-NMD)

Neurorehabilitation
10. September 2020

Joint webinar series



‘How to assess and manage spastic gait in rare diseases?’

Gál Ota

General University Hospital - Prague, Czech Republic

Outline and learning objectives

Outline

- **Taxonomy** of spastic paresis (in like 5 seconds ☺)
- Clinical **assessment** of spastic paresis **in 5 steps**
- Spastic **gait patterns**
- **Case**: basic & advanced assessment & treatment options

Learning objectives

- Focus on the **basics** but always think **out of the box**!

Taxonomy of spastic paresis

Review

The neurophysiology of deforming spastic paresis: A revised taxonomy

Marjolaine Baude [a,*], Jens Bo Nielsen [b], Jean-Michel Gracies [a]

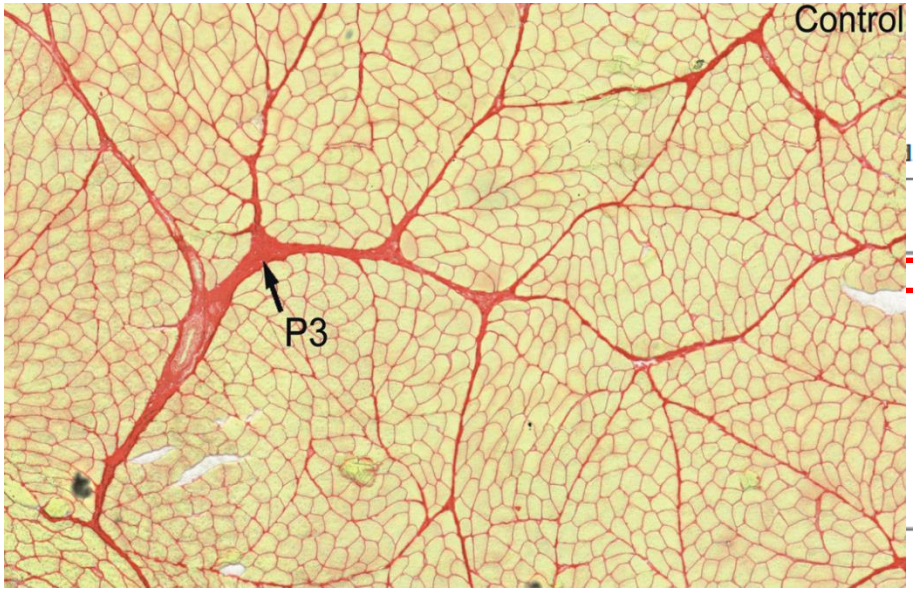
[a] *Service de rééducation neurolocomotrice, EA 7377 BIOTN, laboratoire analyse et restauration du mouvement, université Paris-Est Créteil, hôpitaux universitaires Henri-Mondor, Assistance publique–Hôpitaux de Paris, 51, avenue du Maréchal-de-Lattre-de-Tassigny, 94010 Créteil, France*
[b] *Department of Neuroscience, University of Copenhagen, Blegdamsvej 3, 2200 Copenhagen N, Denmark*

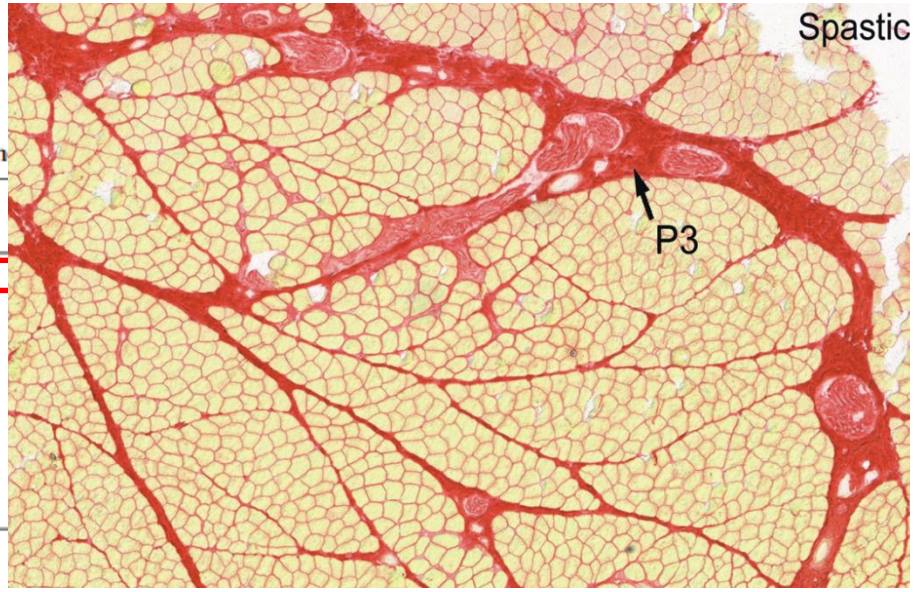
Main features of spastic paresis, with their deforming and disabling properties and their clinical measurability. FRA, flexor reflex afferents.

	Symptom name	Condition of detection	Trigger	Deforming capacity	Disabling level	Measurability at bed side
Muscle disorder	Spastic Myopathy	Rest	N/A	High	High	Estimation possible
Neurological disorder						
Paresis	Stretch-sensitive paresis	Effort	N/A	None	Moderate	No
Muscle overactivity types	Spasticity	Rest	Phasic stretch	None	Low	Yes
	Spastic Dystonia	Rest	None	High	High	No
	Spastic Cocontraction	Effort	Effort directed to agonist	None	High	No
	Extrasegmental cocontraction (synkinesis)	Effort	Effort	Moderate	Moderate	No
	Nociceptive (FRA) spasms	Rest or effort	FRA stimulation	Moderate	High	No

Taxonomy of spastic paresis

- atrophy (hypotrophy and loss of muscle fibres)
- physical shortening (loss of sarcomeres in series)
- ↓ extensibility (accumulation of connective tissue)

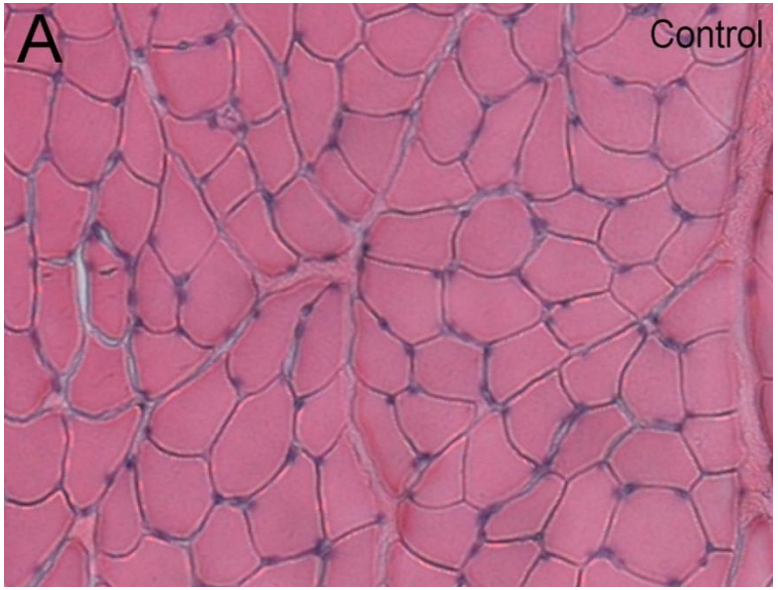


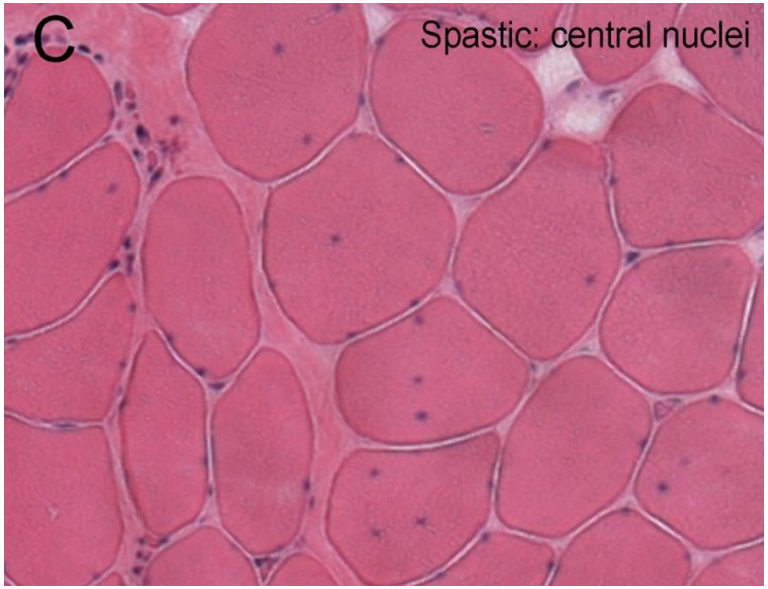


De Bruin 2014

Taxonomy of spastic paresis

- atrophy (hypotrophy and loss of muscle fibres)
- physical shortening (loss of sarcomeres in series)
- ↓ extensibility (accumulation of connective tissue)
- centralization of nuclei?





De Bruin 2014

How to clinically assess spastic paresis?

Update article

Coefficients of impairment in deforming spastic paresis

J.-M. Gracies

Step 1 – Subjective and objective assessment of **function**

Step 2 – **PROM**$_{max}$ (X_{v1})

Step 3 – Angle of **catch** or clonus (X_{v3}) and spasticity grade (Y)

} = **Modified Tardieu Scale**

Step 4 – **AROM**$_{max}$ (X_A)

Step 5 – Amplitude **decrement** (X_{A15}) from X_A in time given, RAM frequency

	Objective	Subjective
Upper extremity	MFS	GSSA
Lower extremity	10MWT, 2MWD	GSSA

Strategy of spastic gait analysis

1. Evaluate the influence of **overactive antagonists** on gait phases in 10MWT (**step 1**)

THEIR NEGATIVE IMPACT IS INCREASED WHEN STRETCHED

(the more it is stretched, the more it co-contracts)

Strategy of spastic gait analysis

1. Evaluate the influence of **overactive antagonists** on gait phases in 10MWT (**step 1**)
2. Go through **steps 2-5** for these antagonists
3. Count the **coefficients** (to be explained later)
4. Relate your **findings from 10MWT to the coefficients**
5. Set priorities with respect to **patient's goals**

Coefficients of impairment

Coefficient of shortening $\frac{(X_N - X_{V1})}{X_N}$ how much shortening (in %) is present

Coefficient of spasticity $\frac{(X_{V1} - X_{V3})}{X_{V1}}$ how much spasticity (in %) is present in the available PROM

Coefficient of weakness $\frac{(X_{V1} - X_A)}{X_{V1}}$ how much weakness (in %) is present in the available PROM

Gracies 2015

Anterior and posterior spastic gait pattern

Anterior pattern

- Insufficient hip extension and early start of swing
- Impairs knee flexion in swing (stiff knee)
- „normal“ step length (paretic side), ↓ speed, swing asymmetry

\+ GCM: ↓ passive dorsiflexion and thus late hip extension

Posterior pattern

- Decreased hip flexion in swing (esp. GM)
- Impaired knee extension in swing (only HAM)
- Decreased step length on paretic side (both GM+HAM)
- GM: compensatory recruitment of RF (hip flexor)

  → imitates ant. pattern but ↓ step length & good late stance!

Gracies 2015

Impact of overactive muscles on gait: triceps

SOLEUS

- First ½ of swing: tripping, slipping; CAVE: anterior pattern!
- In mid stance: knee hyperextension

GASTROCS

- Second ½ of swing: tiptoeing and decreased step length

Gracies 2016

Impact of overactive muscles on gait: adductors

HIP FLEXOR-ADDUCTORS (longus, brevis, pectineus)

- First ½ of swing: scissoring (slow, energy consuming, rel. good balance)

HIP EXTENSOR-ADDUCTORS (magnus, gracilis)

- Second ½ of swing: ↓ base and LL crossing (faster, poor balance)

Impact of overactive muscles on gait: others

TIBIALIS POSTERIOR: foot inversion in swing (dystonia also in stance)

TOE AND BIG TOE FLEXORS: flexion in swing (dystonia also in stance)

LONG BIG TOE EXTENSOR: extension in swing (dystonia also in stance)

Gracies 2016

Case report
(general slides follow)

ITB

Indications	Implant in	Level of evidence
Spastic quadruplegia in CP	Low C / High Th	↓ muscle tone BLE (I) overall ↓ tone (II)
Spastic paraplegia of spinal origin	Mid / Lower Th	↓ tone and spasms (I) ↑ function (III)
Hereditary spastic paraparesis	Mid / Lower Th	↓ spasticity (IV) CAVE: ↓ strength
Secondary generalized dystonia	Lower C / High Th / Intraventricular	(IV)

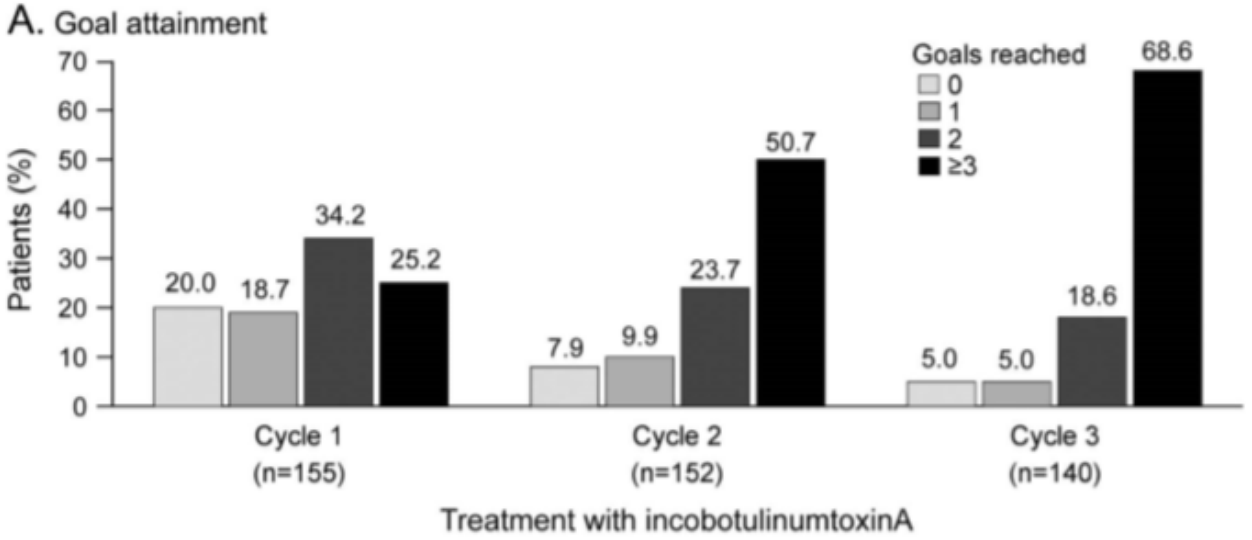
Lake 2019

Complications	Rate	Intervention in response
Infection	3-9,3 % (1) 5-26 % (2)	Remove and treat (infection and withdrawal sy)
Cerebrospinal fluid leak	3,3-4,9 % (2)	Revise wound
Catheter and/or pump malfunction	4-24 % (2)	Revise catheter, repair/replace pump
Severe adverse events	23 % (1)	Improve quality of care

1) Lake 2019, 2) Woolf 2017

How to increase BTX dose?

Cycle 1 (12-16 weeks)	Cycle 2 (12-16 weeks)	Cycle 3* (12-16 weeks)
Maximum of 400 U per limb	Maximum of 600 U per limb	Maximum of 600 U per limb
Total body dose 400 U	Total body dose 600 U	Total body dose 800 U*



Wissel 2017

How to increase BTX dose?

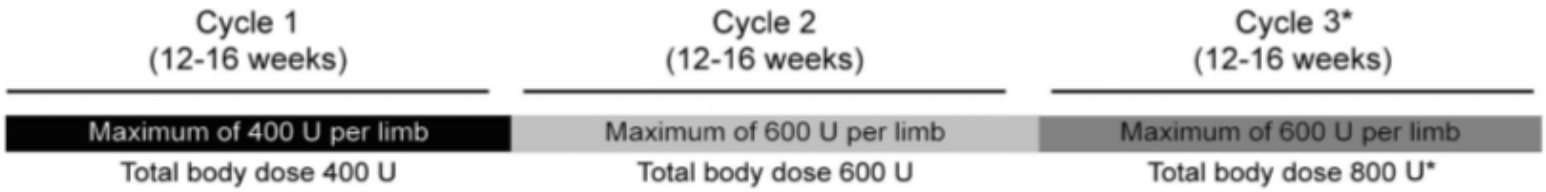
Cycle 1 (12-16 weeks)	Cycle 2 (12-16 weeks)	Cycle 3* (12-16 weeks)
Maximum of 400 U per limb	Maximum of 600 U per limb	Maximum of 600 U per limb
Total body dose 400 U	Total body dose 600 U	Total body dose 800 U*

Table 2 Summary of adverse events by injection cycle

				Cycle 3	
	Overall (n = 155)	Cycle 1 (n = 155)	Cycle 2 (n = 152)	All doses (n = 140)	800 U dose (n = 116)
Any treatment-related AE	17 (11.0)	7 (4.5)	8 (5.3)	4 (2.9)	3 (2.6)
Any AESI	19 (12.3)	6 (3.9)	8 (5.3)	7 (5.0)	6 (5.2)
Any treatment-related AESI[a]	8 (5.2)	2 (1.3)	4 (2.6)	3 (2.1)	3 (2.6)
Any serious AE	17 (11.0)	4 (2.6)	11 (7.2)	3 (2.1)	3 (2.6)
Any treatment-related serious AE	0	0	0	0	0
Any AE leading to discontinuation[b]	5 (3.2)	1 (0.6)	4 (2.6)	0	0
Any treatment-related AE leading to discontinuation	4 (2.6)	1 (0.6)	3 (2.0)	0	0

Wissel 2017

How to increase BTX dose?



- **Increasing improvements** in AS, REPAS, FAC, GAS, DAS, Likert scale
- AE occurrence **was not dependent** on:
  - increasing **dose**
  - **repeated** injections
  - **frequency** of injections every 6 weeks (Evidente 2014, Jankovic 2011)
- Bensmail 2020: effective in **equinovarus**

Wissel 2017

Turbomed

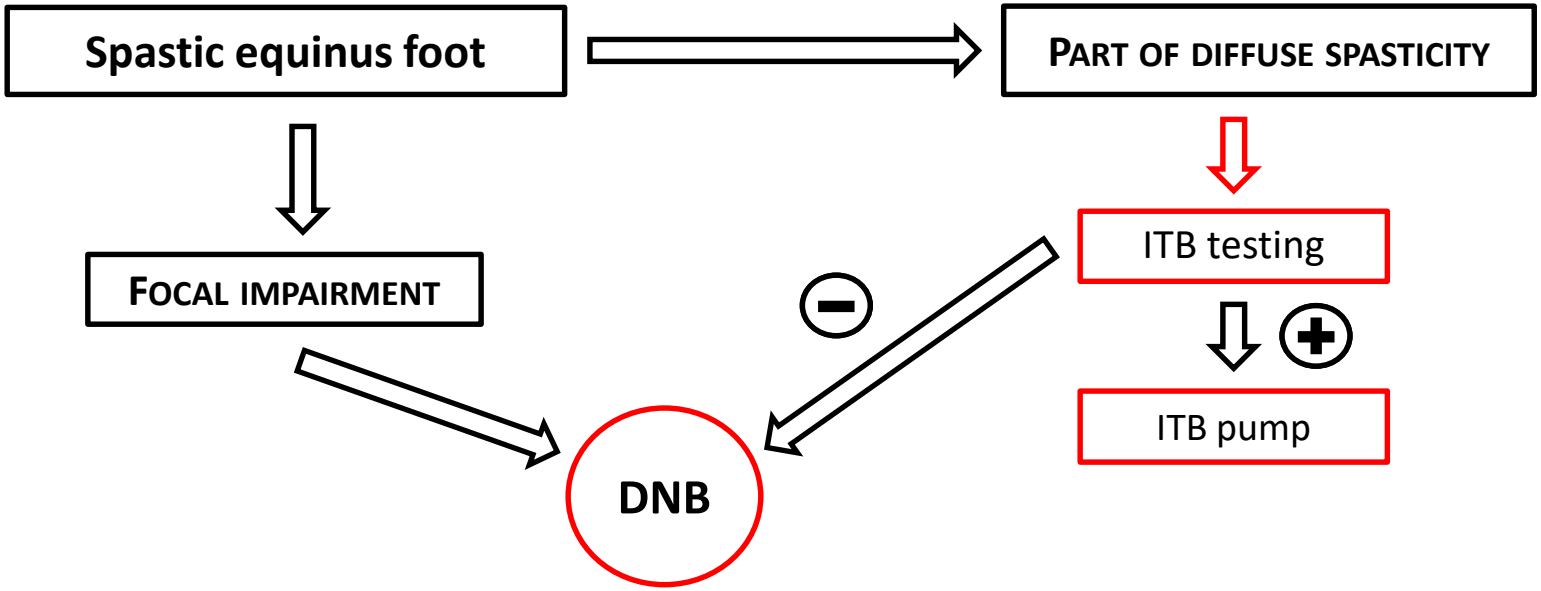
Turbo Med™
ORTHOTICS

Mont-Godinne Guidance Pathway

ASSESSMENT AND TREATMENT OF SPASTIC EQUINOVARUS FOOT AFTER STROKE: GUIDANCE FROM THE MONT-GODINNE INTERDISCIPLINARY GROUP

Thierry DELTOMBE, MD[1], Delphine WAUTIER, MD[2], Philippe DE CLOEDT, MD[2], Michèle FOSTIER, MD[3] and Thierry GUSTIN, MD[4]

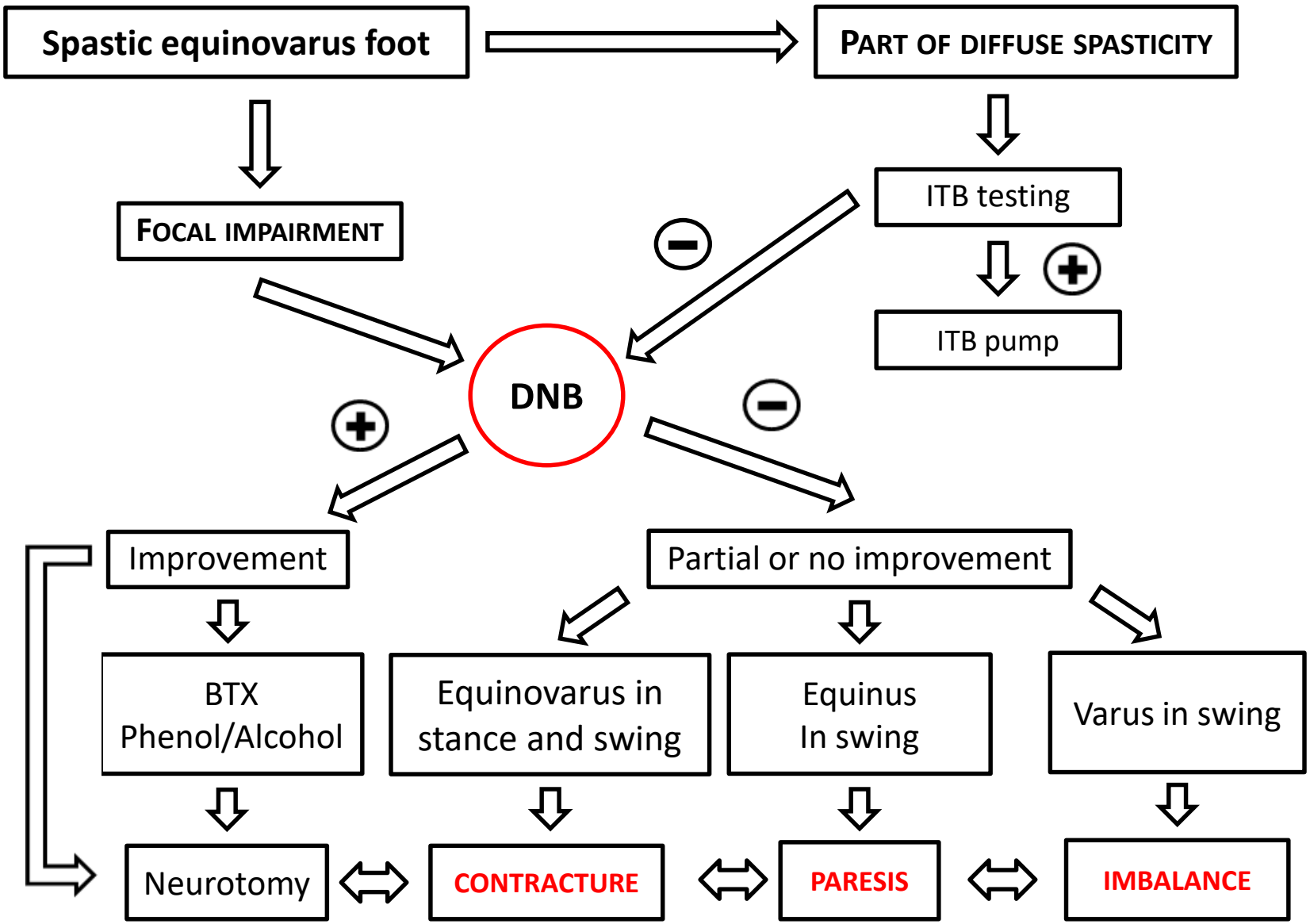
From the Departments of [1]Physical Medicine and Rehabilitation, [2]Orthopaedic Surgery, [3]Anaesthesiology and [4]Neurosurgery, CHU UCL Namur site Mont-Godinne (Université catholique de Louvain), Yvoir, Belgium



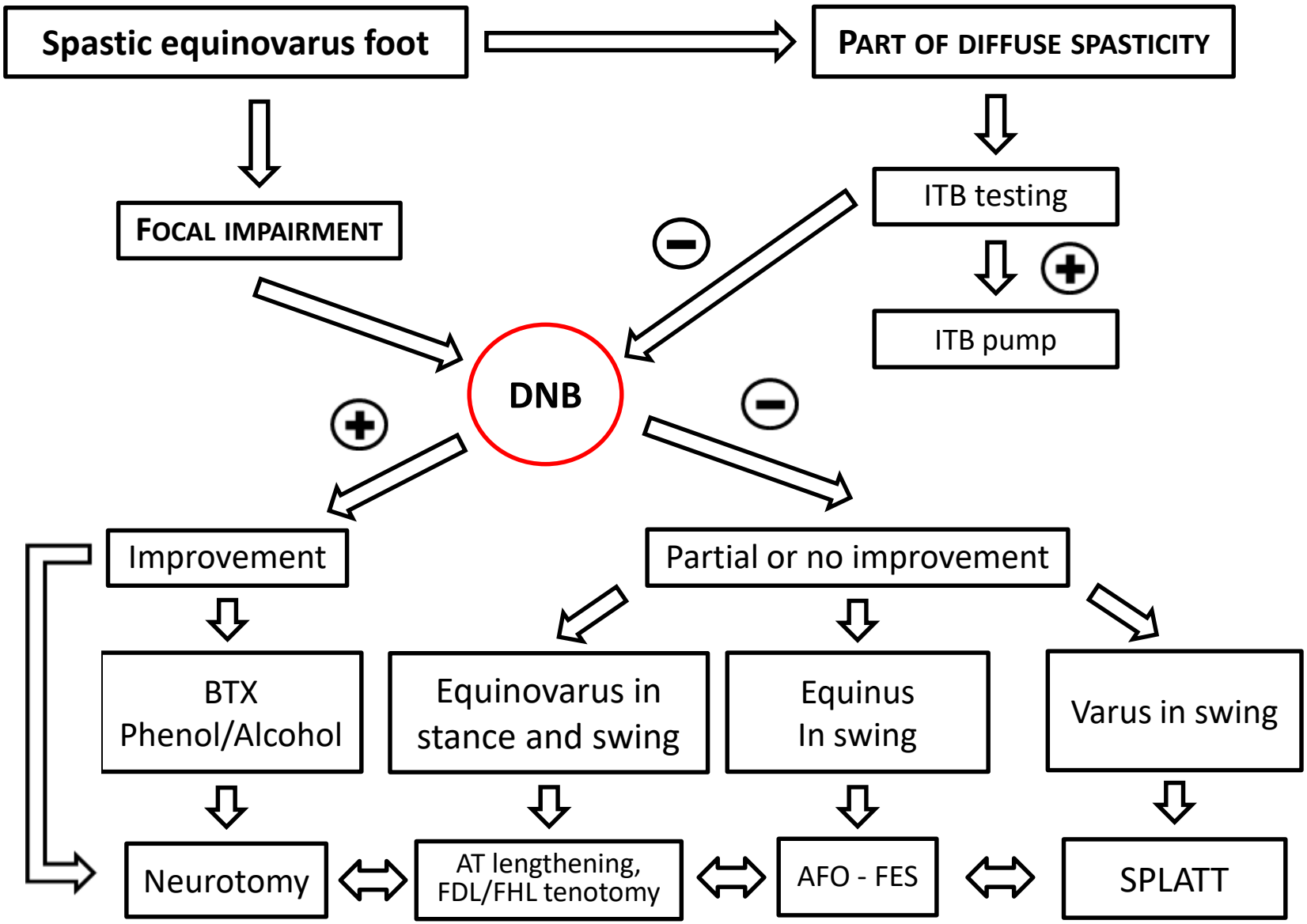
Diagnostic nerve block

- injecting anaesthetic (1-2ml 2% lidocaine) to the nerve to temporarily reduce muscle overactivity
- needle for conduction anaesthesia, stimulator/EMG (H-reflex)
- **SELECTIVE DNB**: motor nerve branch to individual muscles
  ➡ involvement in gait pathology, type and therapy effect
- **NON-SELECTIVE DNB** of tibial nerve (GCM, SOL, TP, FD & H)
  ➡ differentiation of contracture and dystonia

Mont-Godinne Guidance Pathway



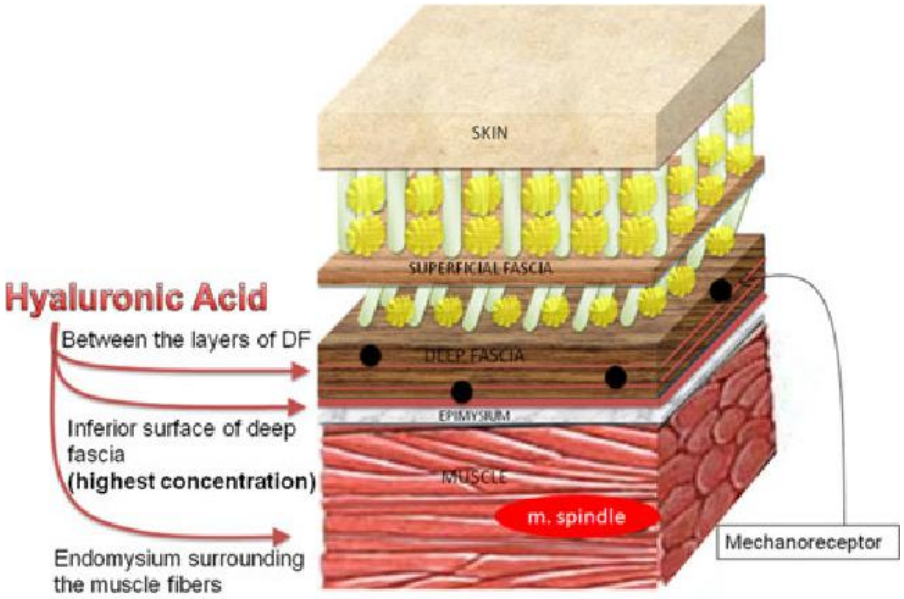
Mont-Godinne Guidance Pathway



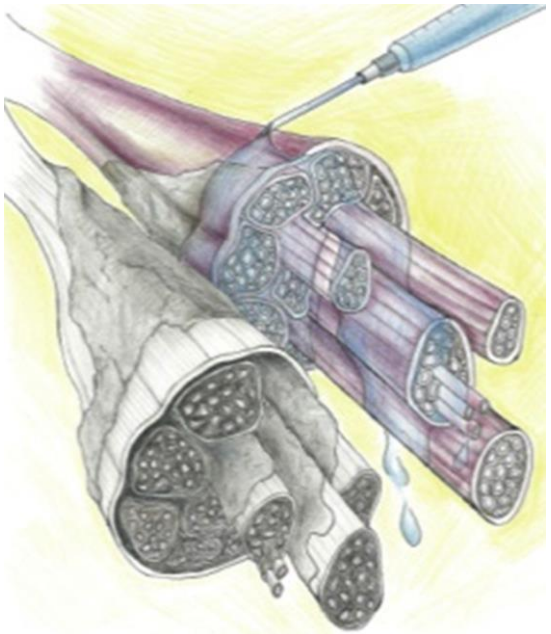
Hyaluronidase as an alternative?

STECCO 2014 (hyaluronan hypothesis)

- Accumulation of HA in a muscle leads to ↑ stiffness



Stecco 2011

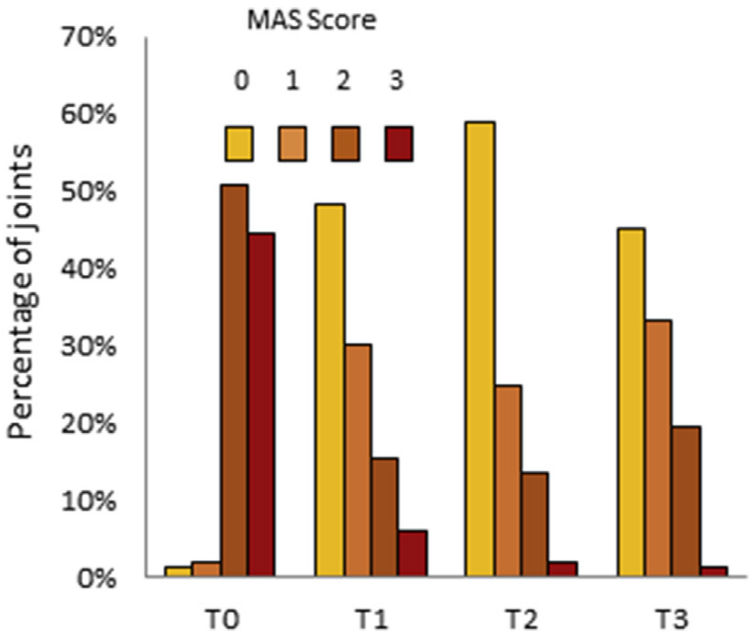


Raghavan 2018

Hyaluronidase as an alternative?

RAGHAVAN 2016 (hyaluronidase injections)

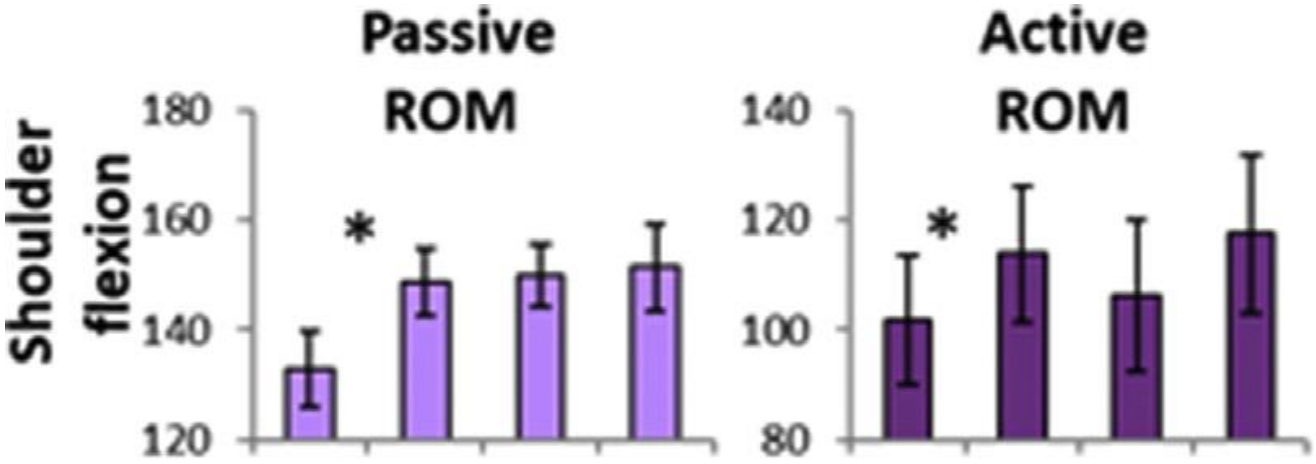
- n=20, UL spasticity (stroke, CP, tumor)
- Assessed after injection, after 4-6W & 3-5M
- ↑ effect on PROM, spasticity (AS) & AROM; no AE



Hyaluronidase as an alternative?

RAGHAVAN 2016 (hyaluronidase injections)

- n=20, UL spasticity (stroke, CP, tumor)
- Assessed after injection, after 4-6W & 3-5M
- ↑ effect on PROM, spasticity (AS) & AROM; no AE



Cannabidiol

- evidence for the **effect on spasticity**:
  - ALS (Riva 2019)
  - MS (inconsistent: Inglet 2020 R, Fiani 2020)

    (x Patti 2020: 43.8% of 1432 PwMS improved in ≥1 SRS)

Conclusion: How to assess gait in RD?

key points

As in spastic paresis of any other aetiology, focus on:

- functional assessment (10MWT, 2MWD)
- shortening, muscle overactivity, paresis (5-SCA)
- patient's goals

Explore advanced assessment options:

- diagnostic nerve block
- ITB testing

Conclusion: How to manage gait in RD?

key points

As in spastic paresis of any other aetiology, focus on:

- Muscle overactivity (BTX, ITB, antispastic drugs)
- Paresis (intensive, specific and regular active training)
- Muscle shortening (stretching)

Explore advanced and novel treatment options:

- Surgery (release, lengthening, transfers, neurotomy, ITB)
- Fascial manipulation and/or hyaluronidase injection
- THC/CBD

Establish an honest relationship with the patient

Co-financed by the Connecting Europe Facility of the European Union

European Reference Network for rare or low prevalence complex diseases
Network Neurological Diseases (ERN-RND)

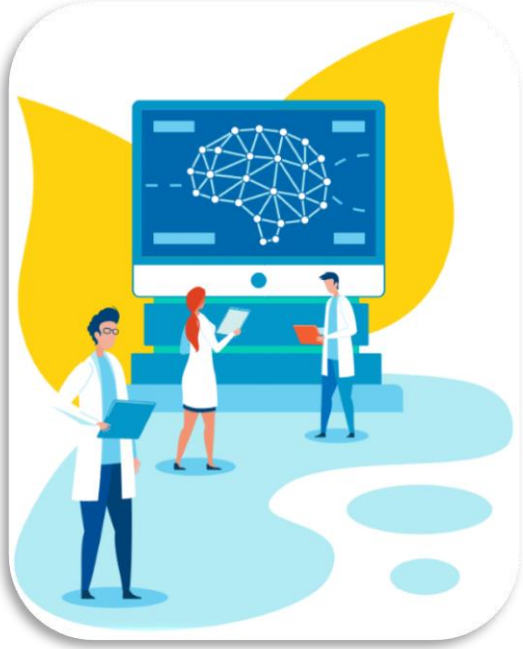
European Reference Network for rare or low prevalence complex diseases
Network Neuromuscular Diseases (ERN EURO-NMD)

Neurorehabilitation
10. September 2020

European Reference Network | Neurological Diseases (ERN-RND)

This webinar has been supported by ERN-RND , which is partly co-funded by the European Union within the framework of the Third Health Programme "ERN-2016 - Framework Partnership Agreement 2017-2021."

Joint webinar series



THANK YOU

Next Webinar: **'A challenge in neurogenetics: Huntington disease in kids'**

15. September 2020, 15-16h CET