Scandinavia goes St. Gallen

Repetive Nerve Stimulation

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Kantonsspital
St.Gallen

Repetitive nerve stimulation (RNS) for neuromuscular transmission diseases

- RNS is a commonly used method to evaluate NM transmission in myasthenia gravis and LEMS

- While RNS is a straight-forward technique, several aspects have to be considered to improve quality of assessments and sensitivity

  - muscle selection
  - Optimal stimulation
  - Electrode position
  - Joint position/muscle contraction
  - Temperature
  - findings

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Repetitive nerve stimulation: muscle selection

- Many muscles can be measured to increase sensitivity, but a "good choice" will decrease efforts of examiners and burden to patients

- In generalized Myasthenia gravis (MG), proximal muscles are mostly involved clinically and in consequence electrophysiologically

  - E.g. trapezius, deltoid, anconeus muscle
    → we prefer anconeus, as it is easily measured, no relevant muscle contraction (like trapezius or deltoid), easily activated and with low discomfort due to stimulation

  - RNS is more sensitive in generalized MG (53-89%) than in ocular MG (20%-67%) (Zinman et al., 2006;Bou Ali et al., 2017)

  - Rarely positive e.g. in small hand muscles (~19%), mostly normal in MuSK-MG

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Repetitive nerve stimulation: facial muscles

- Facial muscles are often considered to by doubtfully by patients (and examiners), but easily done. However, sensitivity is relatively poor

- generalized MG: M. frontalis/nasalis 46%/35%

- ocular MG: M. frontalis/nasalis only 15%/19% Abraham 2016

- In MuSK myasthenia, frequently normal in limb muscles
  → orbicularis oris muscle is also feasible

- Sensitivity of orbicularis oculi? conflicting publications

  case example MuSK-MG

MUSCLE & NERVE

Clinical Research

Repetitive nerve stimulation cutoff values for the diagnosis of myasthenia gravis

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ABSTRACT

Introduction

Repetitive nerve stimulation (RNS) showing ≥ 10% decrement is considered the cutoff for myasthenia gravis (MG), but this has never been validated. The objective of this study was to find an optimal validated cutoff value for decrement on RNS. *Methods*: We performed retrospective chart review of patients who had electrophysiological assessment for possible MG from 2013 to 2015. *Results*: A total of 122 patients with MG and 182 controls were identified. RNS sensitivities for generalized and ocular MG using the traditional ≥10% cutoff value were 46% and 15%, respectively, for frontalis recordings, and 35% and 19%, respectively, for nasalis recordings. Using a decrement cutoff value of 7% for frontalis and 8% for nasalis increased the sensitivities by 6–11%, with specificities of 95–96%. *Conclusions*: For RNS in facial muscles, we suggest a cutoff value of 7–8%, which increases test sensitivity by 6–11%, while preserving high specificity for the diagnosis of MG. Muscle Nerve, 2016 Muscle Nerve **55**: 166–170, 2017
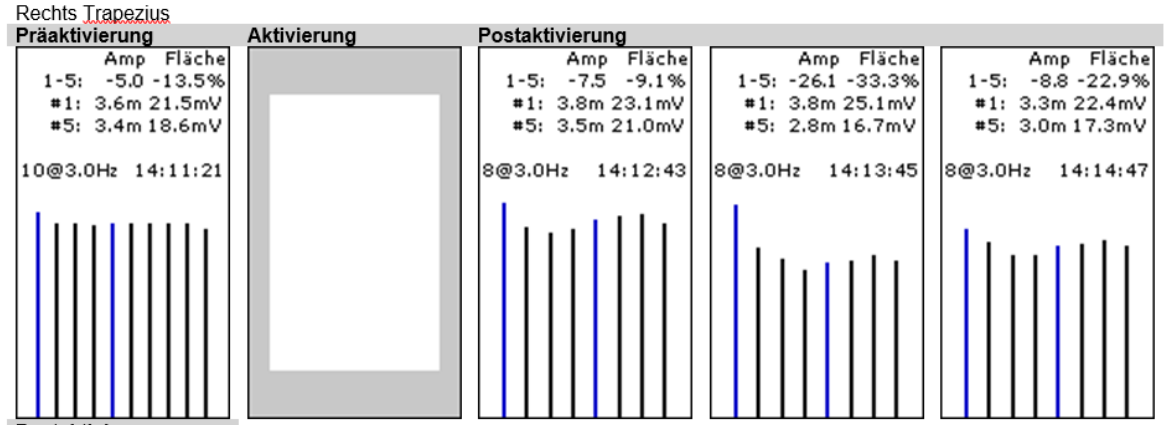
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Repetitive nerve stimulation: facial muscles (case 1)

- Trapezius muscle 2008
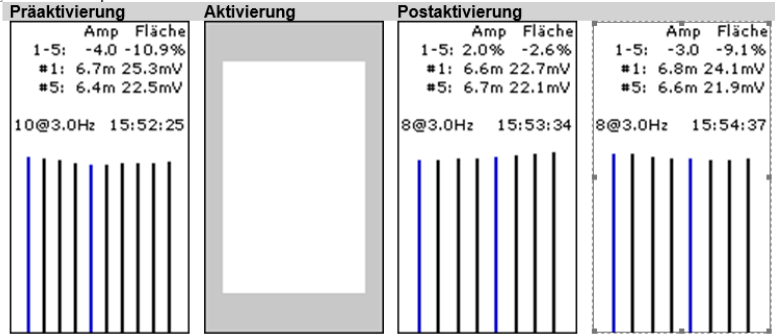→ Fatigue after 2 min
- SFEMG pathological

Single Fiber EMG

Rechts Orbicularis oculi
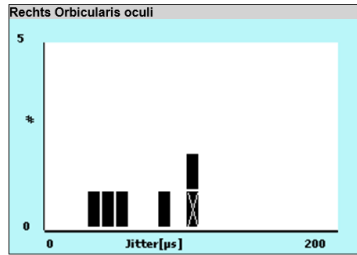
Rechts Orbicularis oculi	Jitter us	MIDI ms	Block
1 (1)	37.1	238	☐
2 (2)	109	29.5	☑
3 (2)	50.3	29.5	☐
4 (3)	236	60.5	☑
5 (3)	85.3	60.5	☐
6 (4)	45.1	36.3	☐
7 (5)			
8 (6)	101	43.3	☐
9 (7)			
Mittelwert	94.7	71.1	

Rechts Trapezius

Präaktivierung	Aktivierung	Postaktivierung
Amp Fläche 1-5: -5.0 -13.5% #1: 3.6m 21.5mV #5: 3.4m 18.6mV 10@3.0Hz 14:11:21		Amp Fläche 1-5: -7.5 -9.1% #1: 3.8m 23.1mV #5: 3.5m 21.0mV 8@3.0Hz 14:12:43

Amp Fläche
1-5: -26.1 -33.3%
#1: 3.8m 25.1mV
#5: 2.8m 16.7mV

8@3.0Hz 14:13:45

Amp Fläche
1-5: -8.8 -22.9%
#1: 3.3m 22.4mV
#5: 3.0m 17.3mV

8@3.0Hz 14:14:47

Postaktivierung

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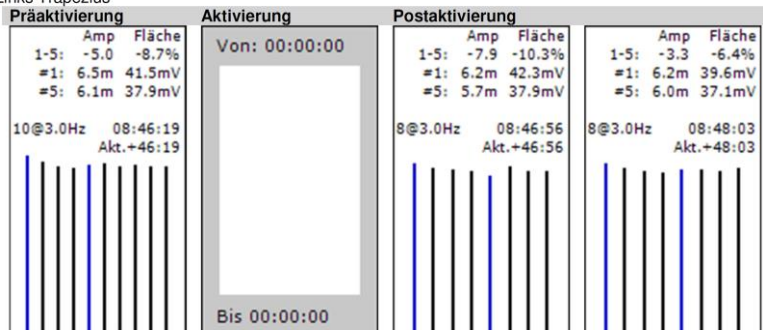- APB, nasalis and orbicularis **oris** muscles 2009
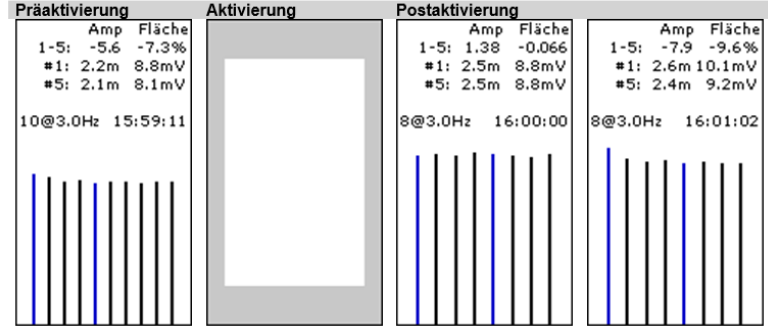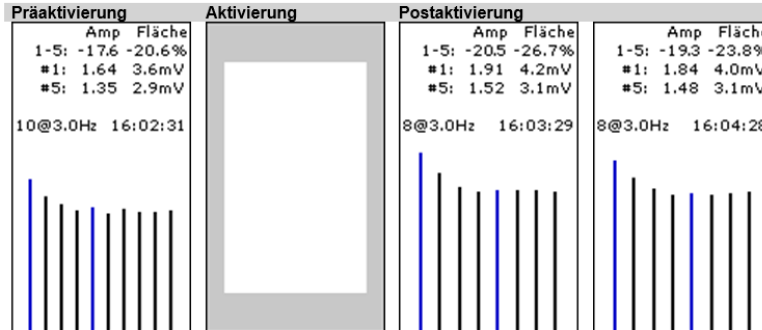
Remark: M. nasalis is a *respiratory muscle* → can be spared in MuSK myasthenia
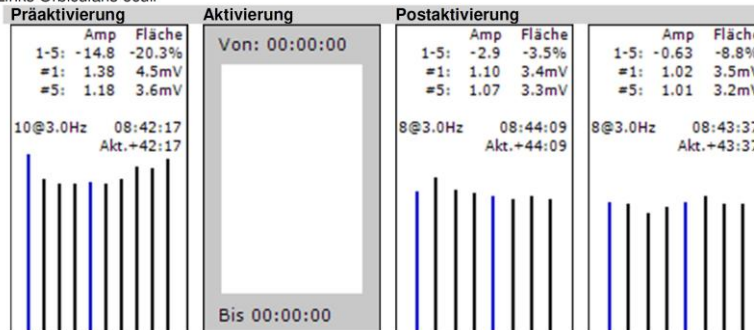
Repetitive nerve stimulation: ocular MG (case 2)

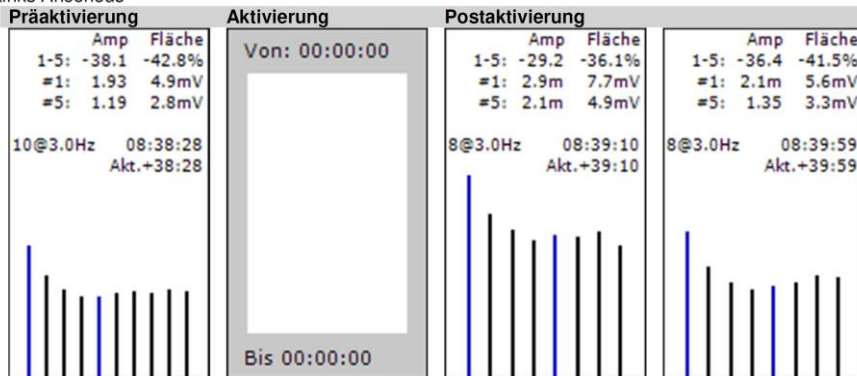- Trapecius, orbicularis **oculi** and anconeus muscles 12/2023

Remark: consider M. anconeus as a sensitive muscle also in ocular MG

Repetitive nerve stimulation:nasalis muscle in MuSK

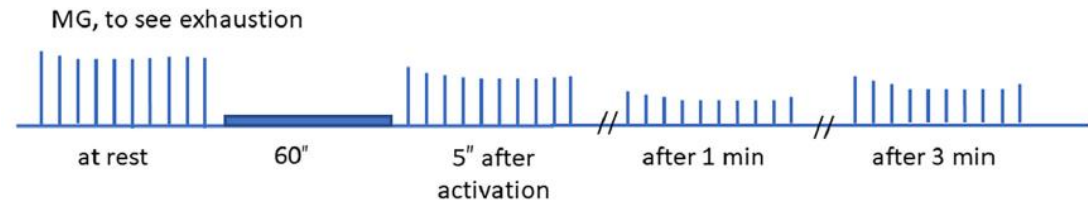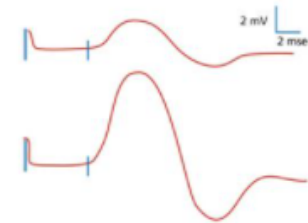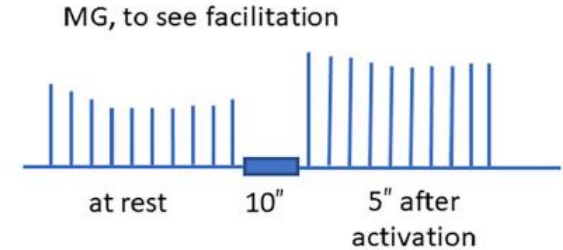Remark: M. nasalis is a *respiratory muscle* → can be spared in MuSK myasthenia

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Repetitive nerve stimulation: joint position/muscle contraction and temperature

- Pitfall: shortening of muscles my electrical stimulation or active movement can move the electrode or muscle belly beneath the electrode, resulting in different CMAP shapes → turn speaker on to detect voluntary activation. Joint position should be stable throughout the measurement

- Sudden or irregular variations between responses are most likely artificial

- Decrement in MG is less when the muscle is cold (inhibition of acetylcholine-esterase like in the ice-test in MG) and sometimes decrement is only seen in a warm muscle and is even higher when the muscle is heated to e.g. 42 °C (not in normal muscles)
  Rutkove et al., 1998

- In LEMS, resting CMAP is much smaller when the muscle is warm

- Warm distal limb muscles if they are cold (if you really want to test them). Warming of proximal or facial muscles is not needed

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Repetitive nerve stimulation: findings in different conditions

- In typical MG, decrement > 10%, often greatest at the 4th or 5th stimulation 3-5Hz
- Sometimes return of amplitude in later stimulations (U-shape or "saddle-shape")
- Often facilitation can bee seen after 10' of maximal contraction
- Alternatively, compare a simple CMAP elicited directly after activation with a resting pre-activation CMAP

- Exhaustion can unmask decrement after longer activation for 60' up to 3 min afterwards

MG, to see facilitation

at rest   10"   5" after activation

2 mV
2 mse

MG, to see exhaustion

at rest   60"   5" after activation   after 1 min   after 3 min

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Comments? Suggestions? Questions…?