

Autoimmun csatornabetegségek és dysimmun neuropathiák elektrofiziológiai diagnózisa

Nota bene: **Nincs** elektrofiziológiai diagnózis, **klinikai** diagnózis van!
(pl.: GBS ENG-je lehet negatív, AMSAN ENG-je kb. mint alkoholos polyneuropathia)

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Áttekintés

Autoimmun csatornabetegségek:

Myasthaenia gravis (MG)	<i>RNS, SFEMG</i>
Lambert-Eaton szindróma (LEMS)	<i>RNS</i>
Isaac's szindróma	<i>EMG - neuromyotónia</i>

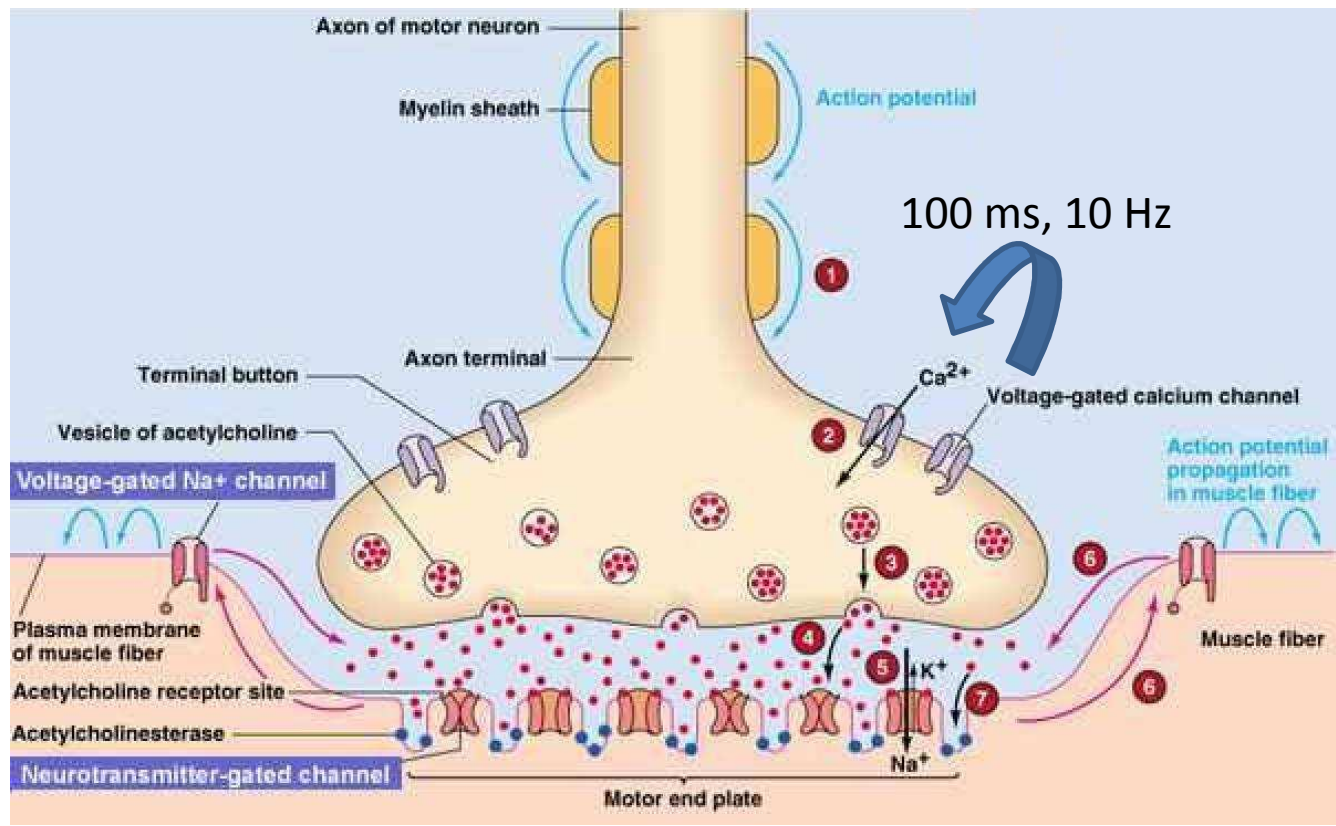
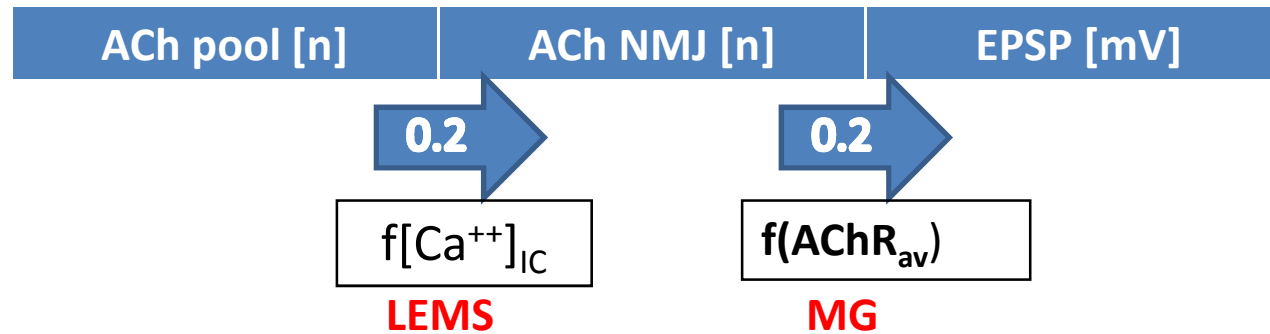
Dysimmun neuropathiák

Guillain-Barre (GBS) csoport

klasszikus GBS	<i>DML, CV, TempDisp</i>
AMSAN	<i>SensMot axon</i>
AMAN	<i>Mot axon</i>
Miller-Fisher szindróma	

CIDP	<i>DML, CV, TemDisp, CondBlokk</i>
DADS	<i>Sens demyelin</i>
MMN	<i>Mot TempDisp, CondBlokk</i>
MADSAM	<i>Mot TempDisp, CondBlokk, sens demyelin</i>

A neuromusculáris junkció (NMJ) elektrofiziológiai modellje



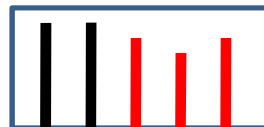
A neuromusculáris junkció (NMJ) elektrofiziológiai modellje – 3 Hz

ACh pool [n]	ACh NMJ [n]	EPSP [mV]
1000	200	40
800	160	32
640	128	25.6
512	102	20.4
640	128	25.6



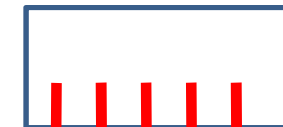
Pool 1 1000 ves
 Pool 2 10 000 ves
 Pool 3 100 000 ves

Myasthaenia gravis



ACh pool [n]	ACh NMJ [n]	EPSP [mV]
1000	200	20
800	160	16
640	128	13
512	102	10
640	128	13

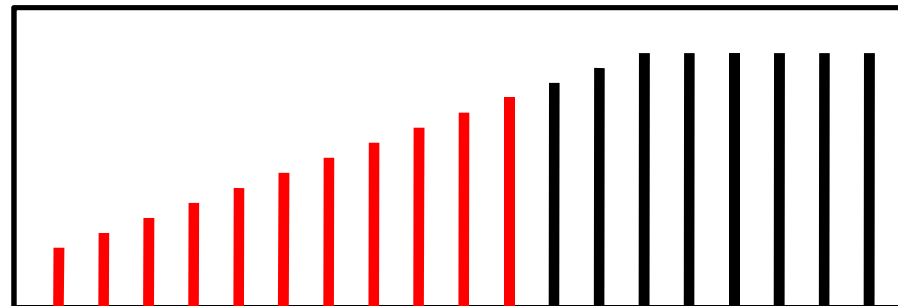
Lambert-Eaton



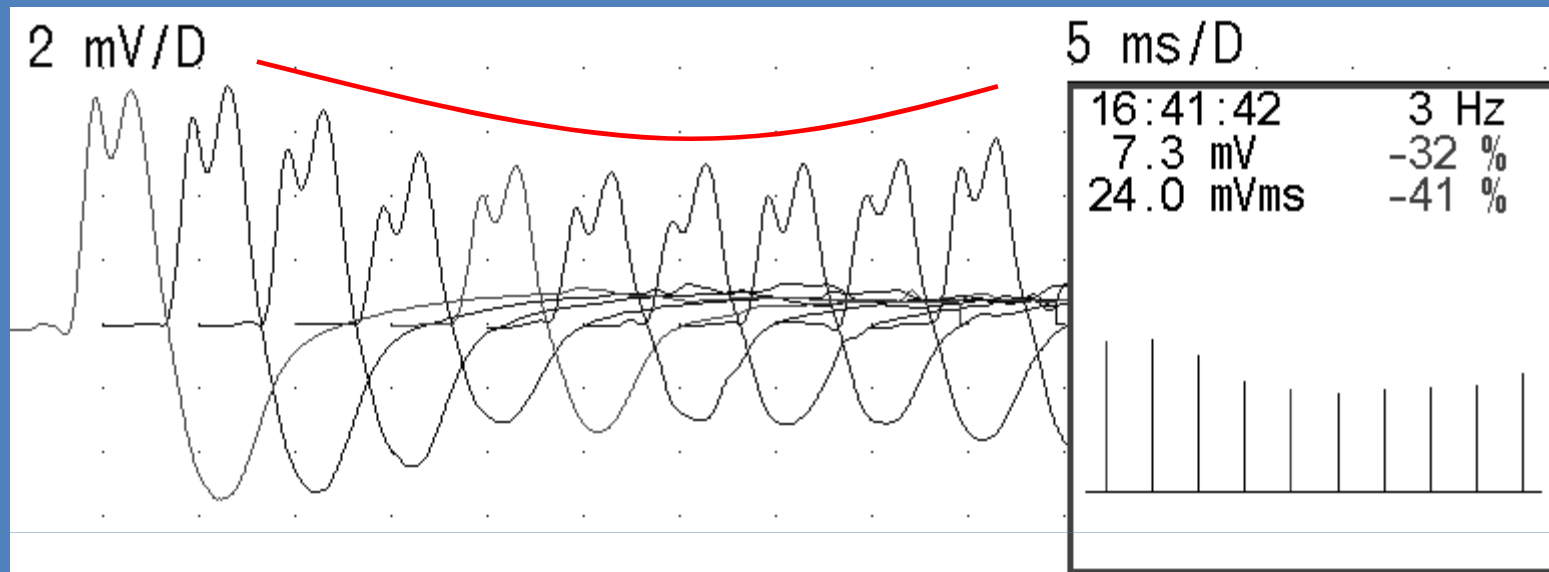
ACh pool [n]	ACh NMJ [n]	EPSP [mV]
1000	20	4
980	16	3.2
964	13	2.6
951	10	2
964	13	2.6

A neuromusculáris junkció (NMJ) elektrofiziológiai modellje – 50 Hz (LEMS)

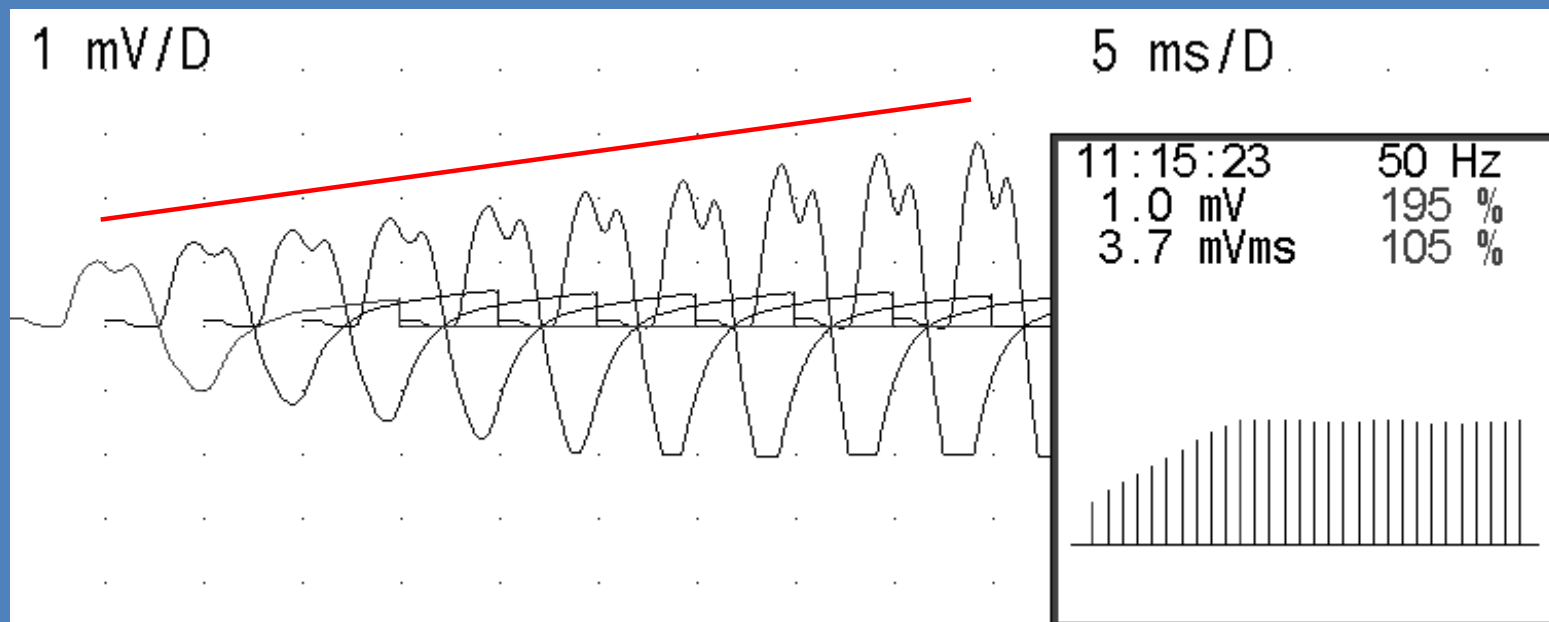
ACh pool [n]		ACh NMJ [n]		EPSP [mV]
1000	0.02	20	0.2	4
980	0.03	29	0.2	5.8
951	0.04	38	0.2	7.6
913	0.05	45	0.2	9
868	0.06	52	0.2	10.4
816	0.07	57	0.2	11.4
....	0.2



Sorozatingerlés

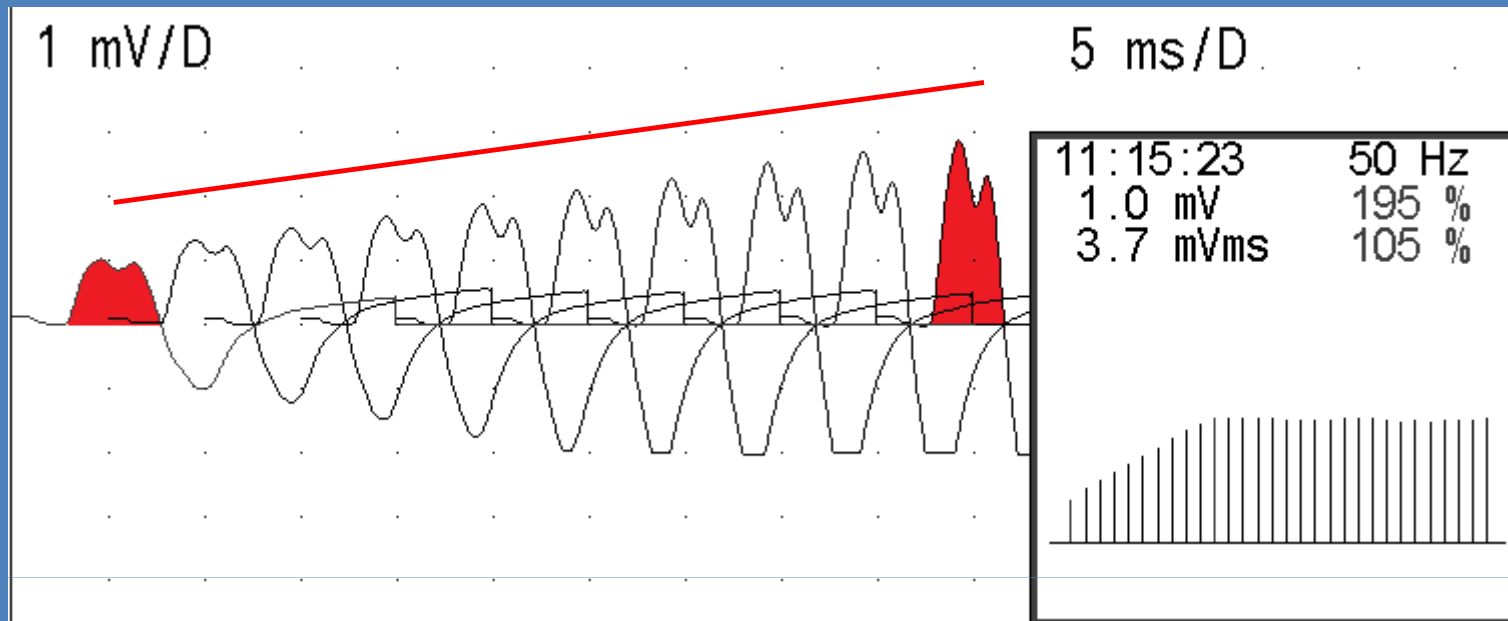


3 Hz
MG

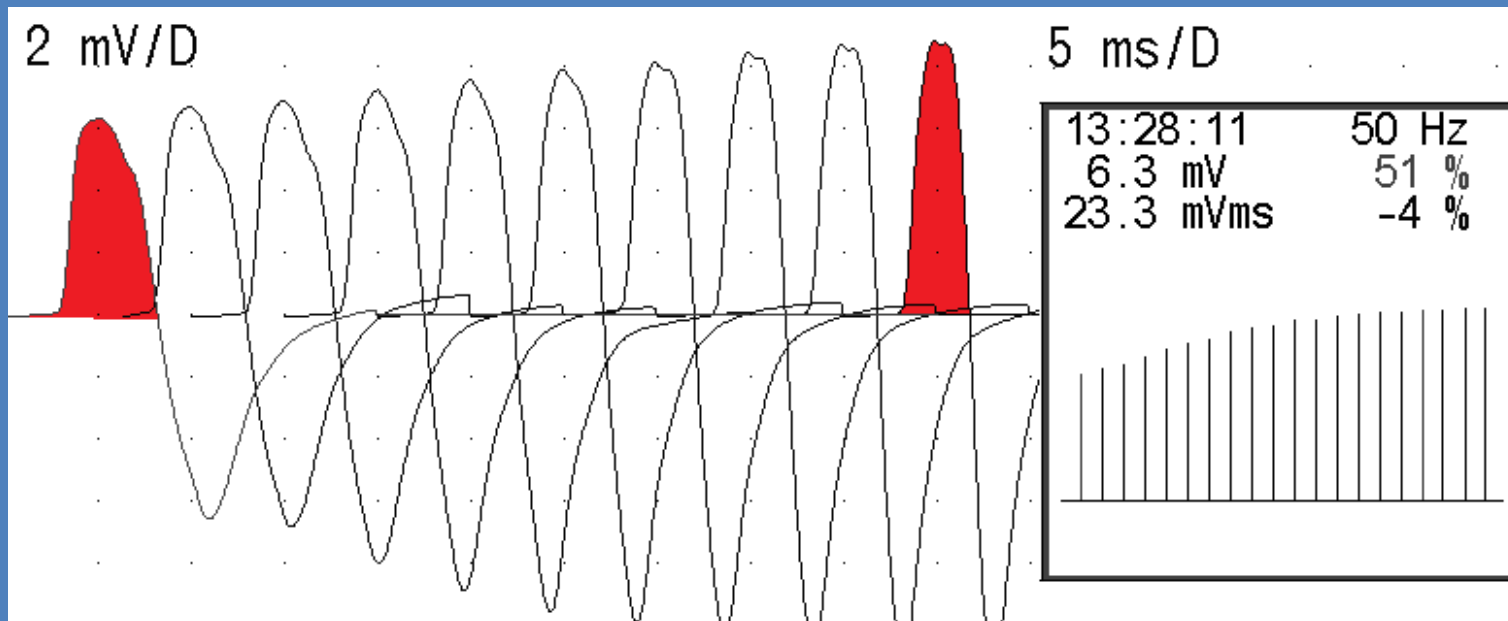


50 Hz
LEMS

Sorozatingerlés, pseudofacilitáció



50 Hz
LEMS



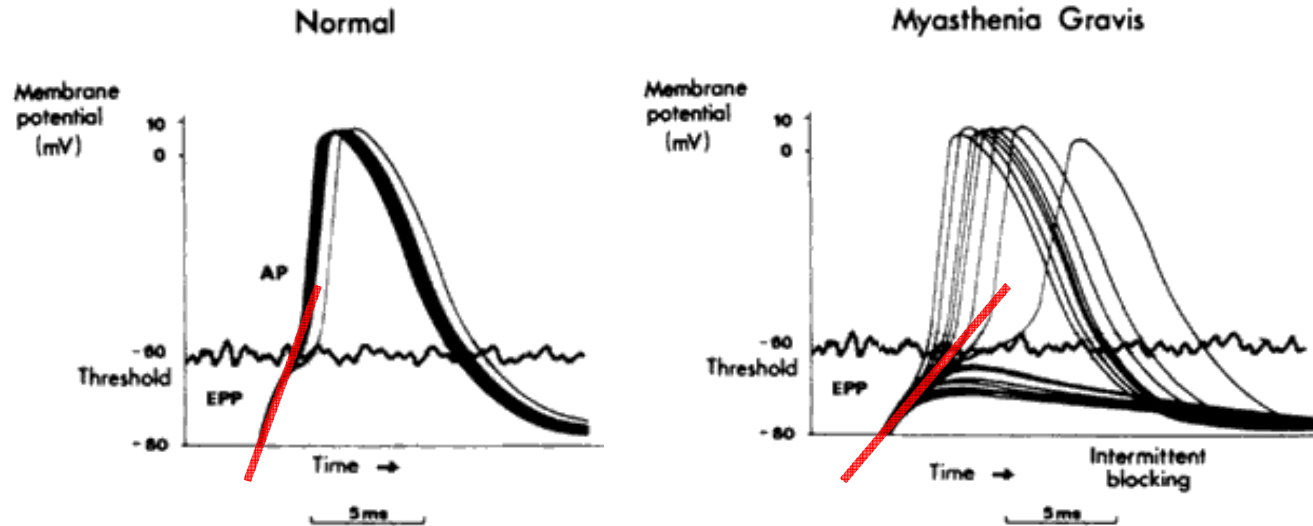
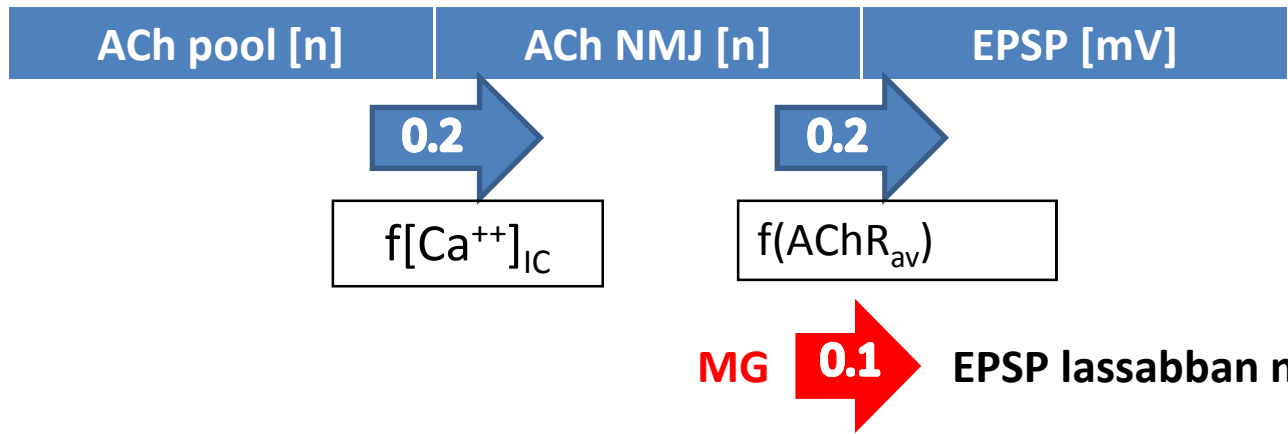
Pseudo-
facilitáció

Ingerületből izopotenciál...

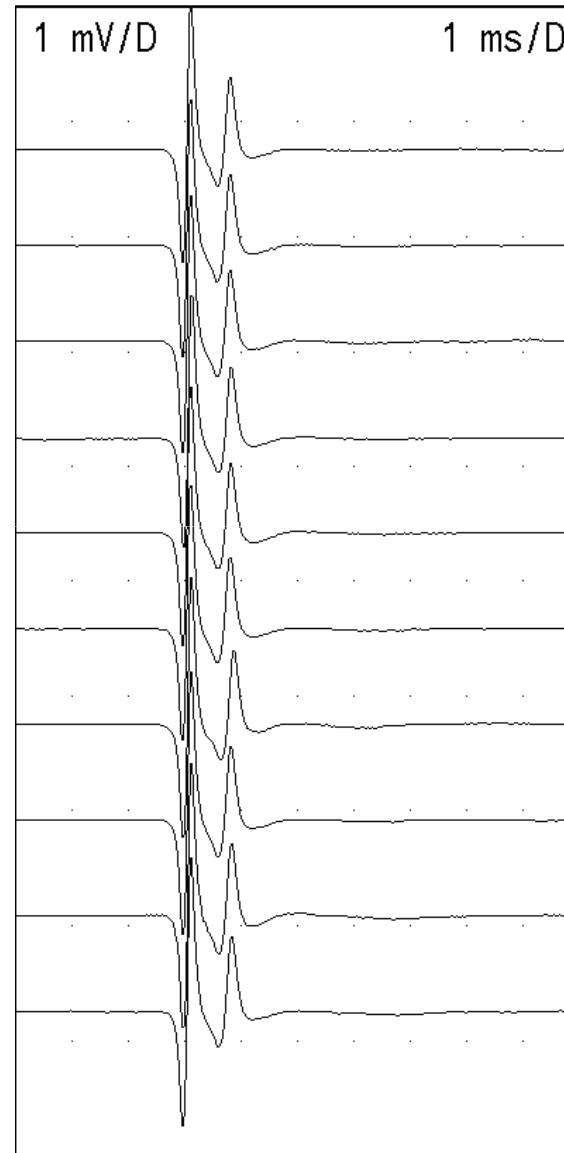
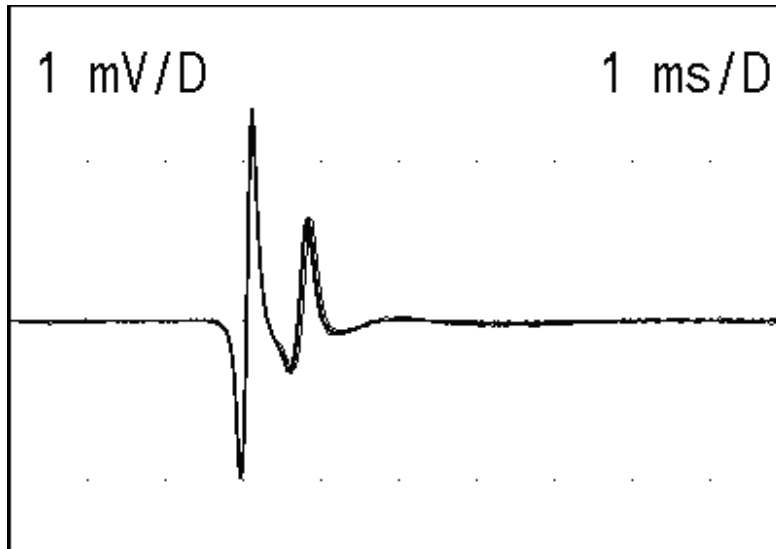
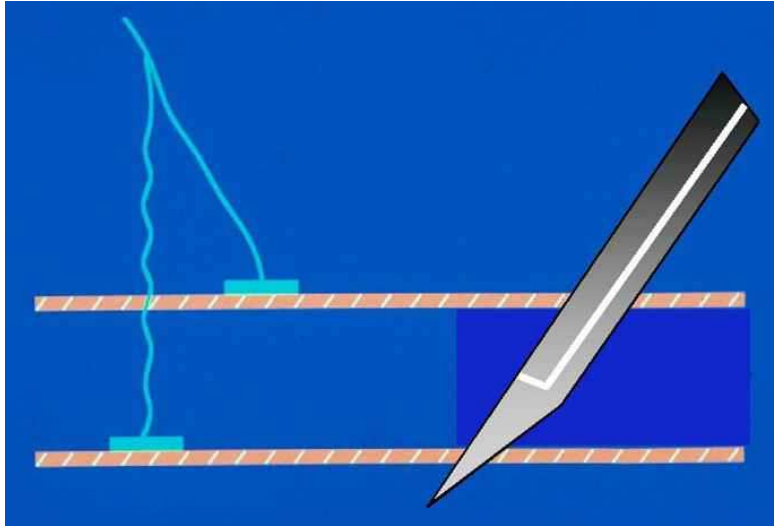
igen/nem **RNS**

mikor? **SFEMG**

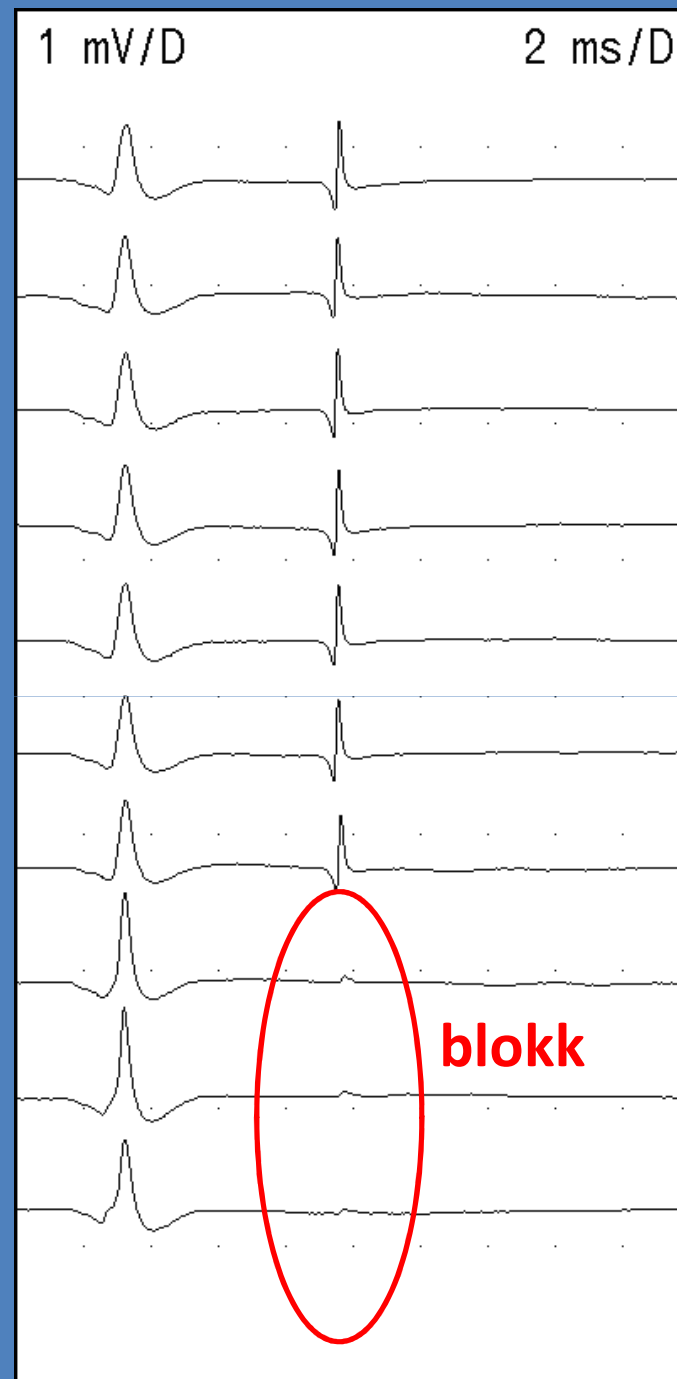
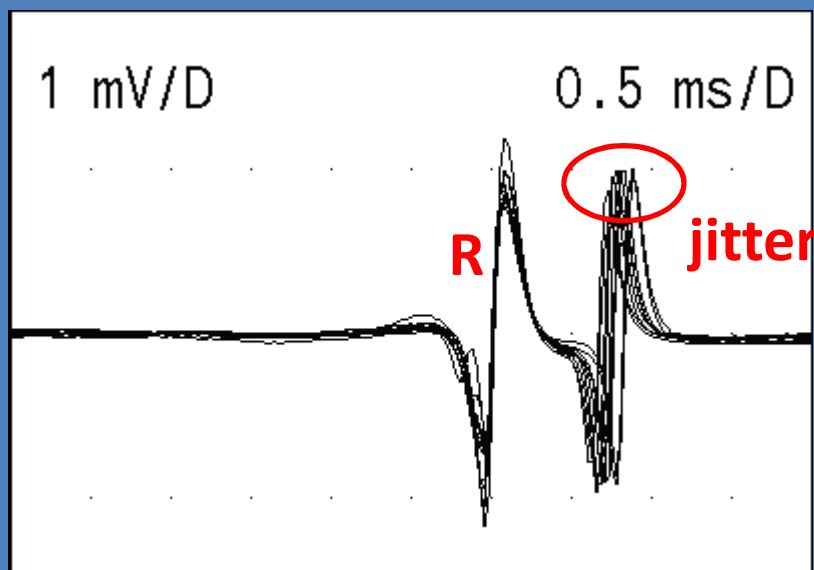
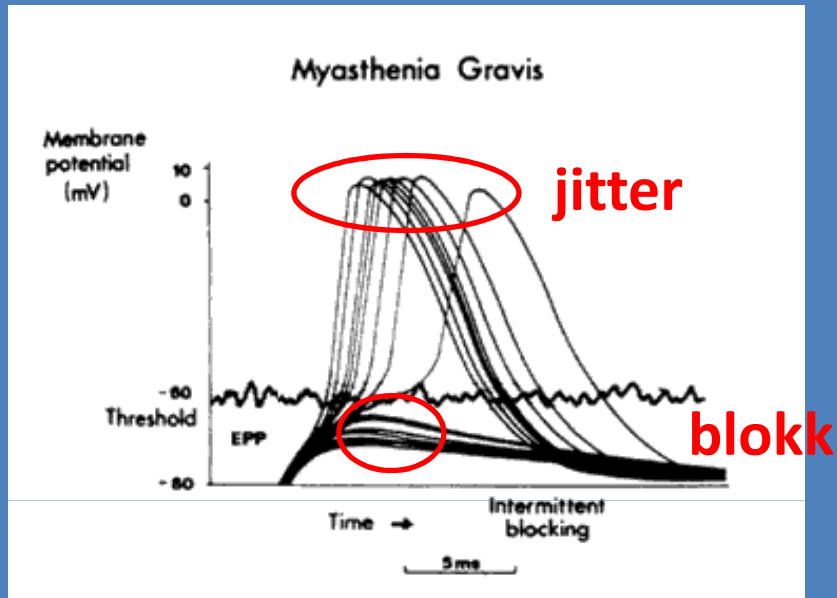
Single fiber EMG



Single fiber EMG



SFEMG; jitter&blokk



Neuromyotónia

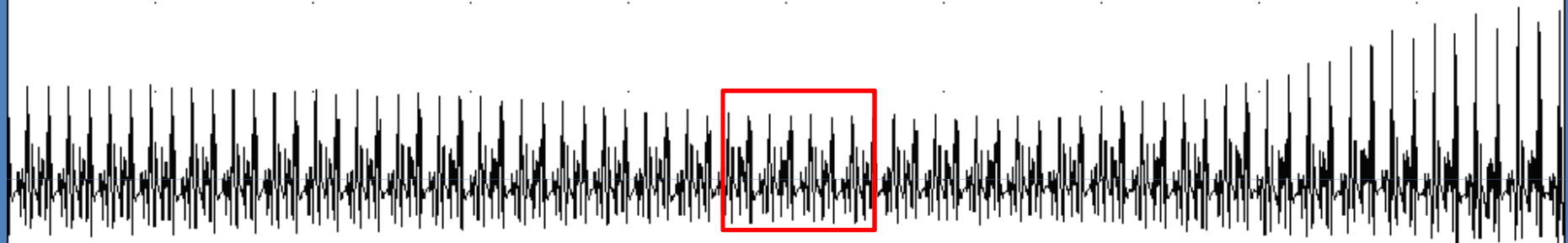
Axonterminálison a VGKC az egyetlen, ami a depolarizáció ellen hat



Folyamatos izomaktivitás (nem izomrost, de nem is mindig MUAP)

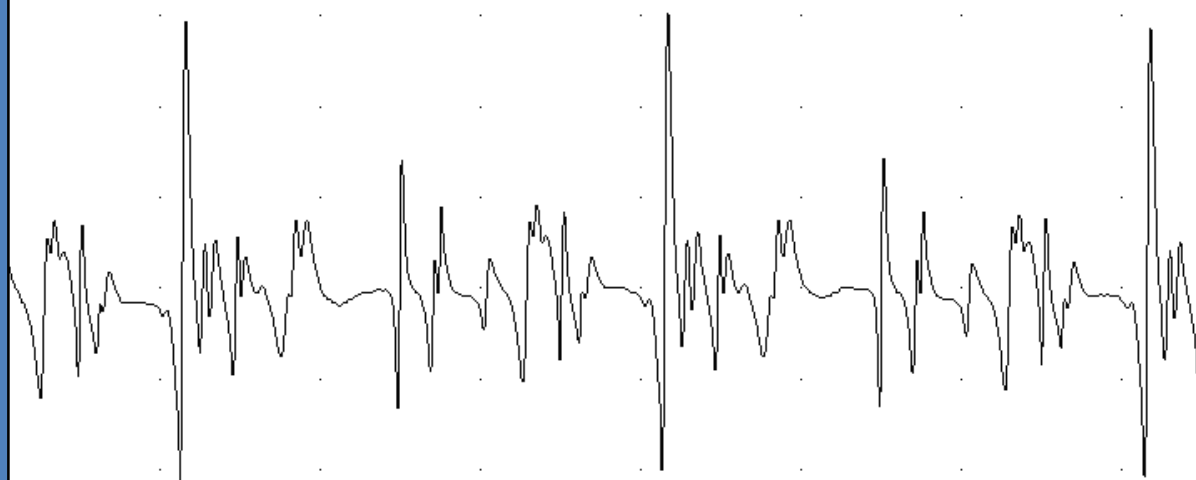
0.2 mV/D

100 ms/D

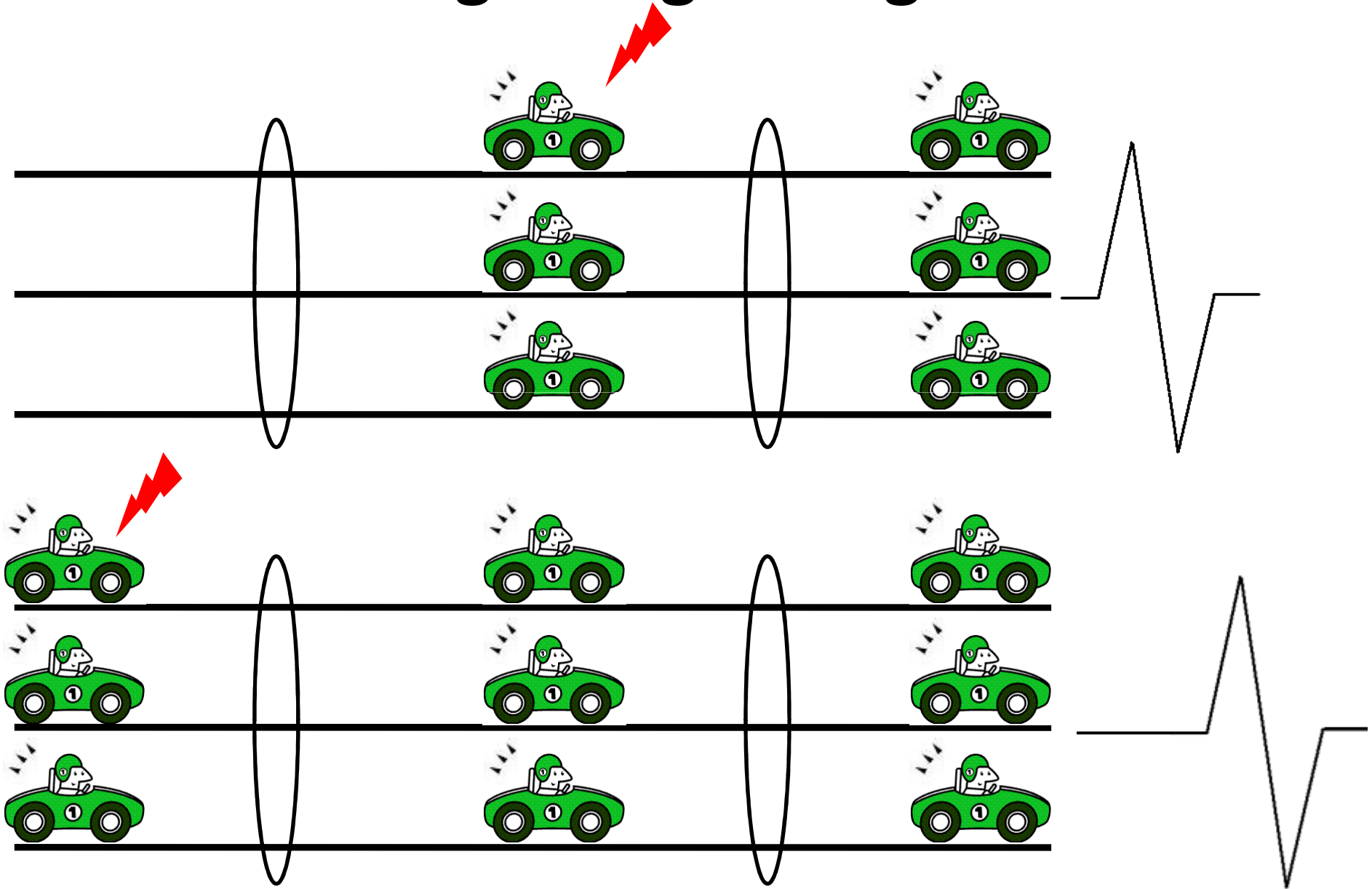


0.2 mV/D

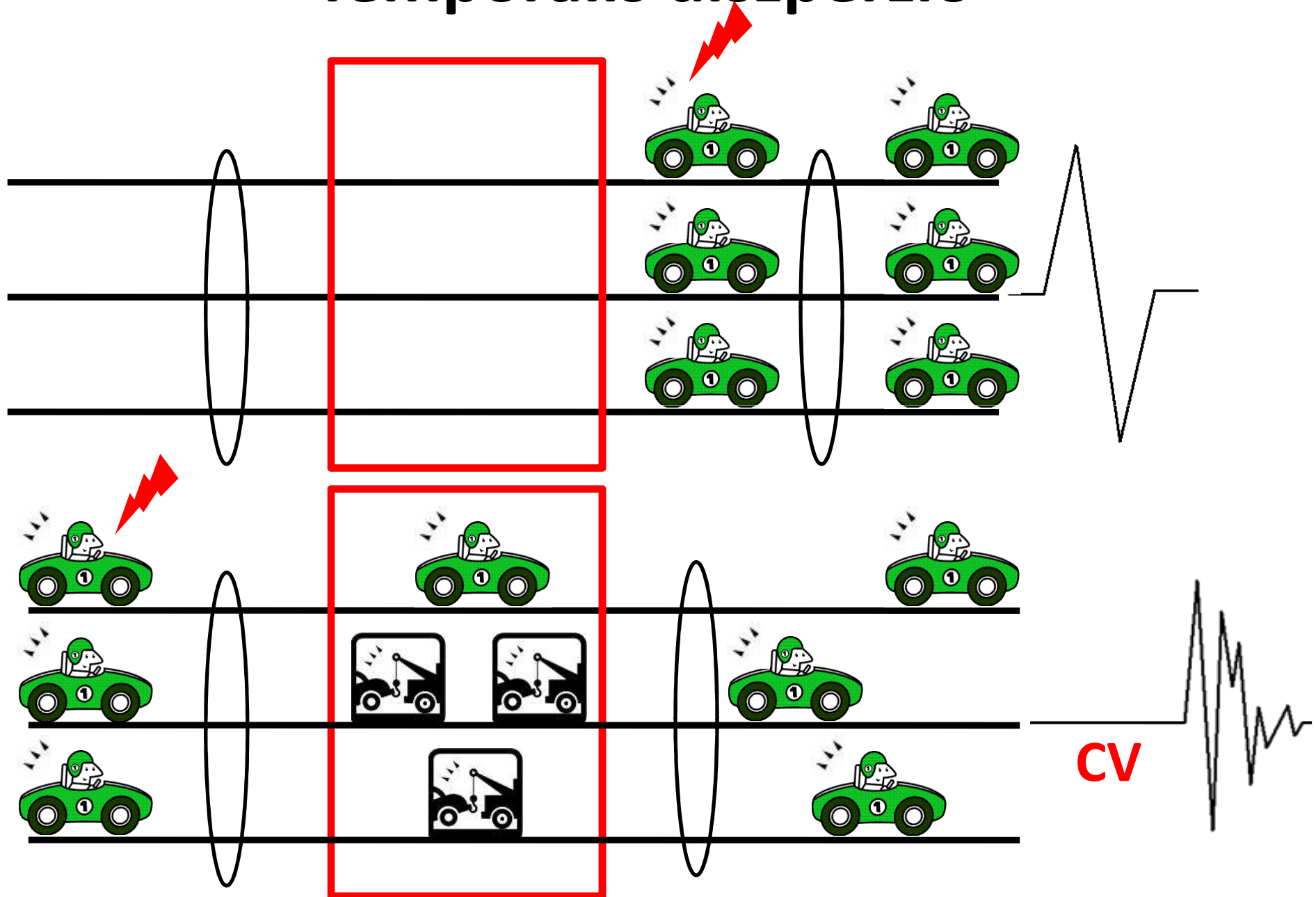
10 ms/D



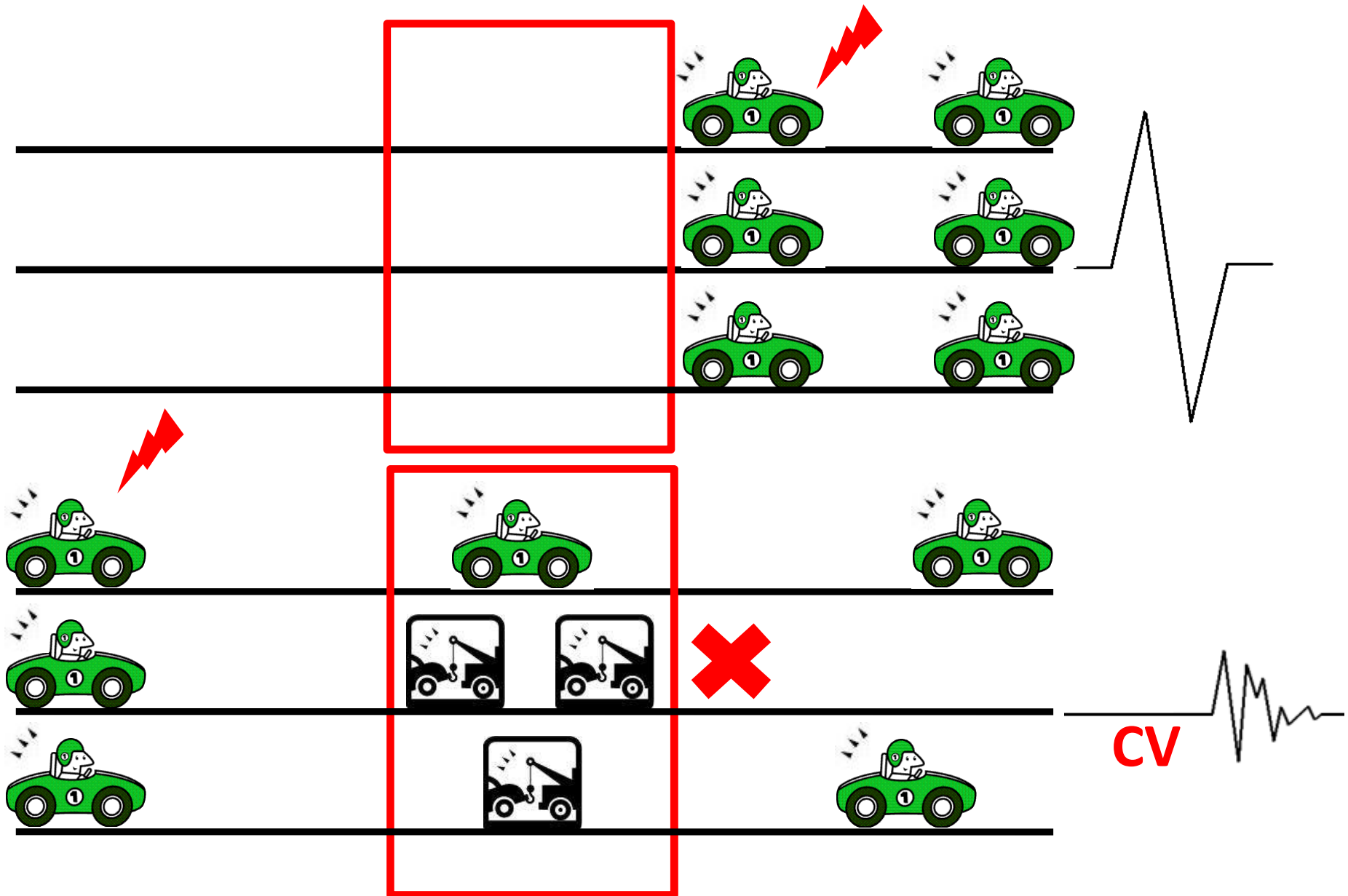
Egészséges ideg



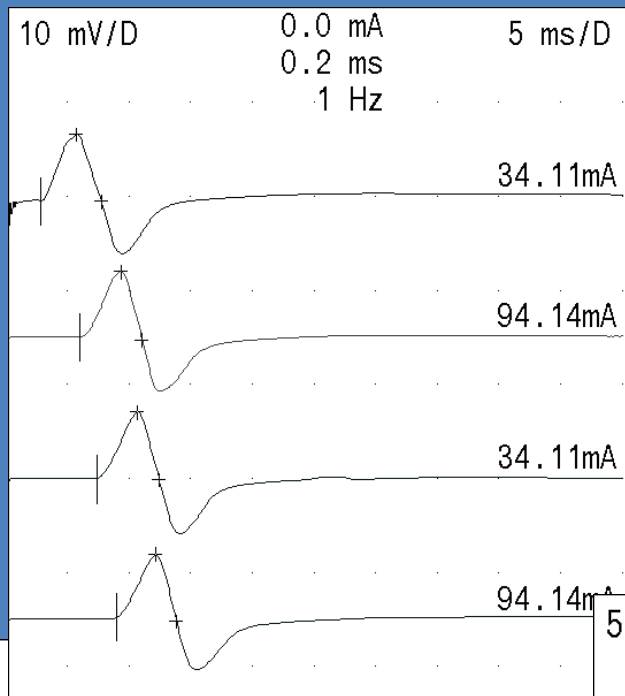
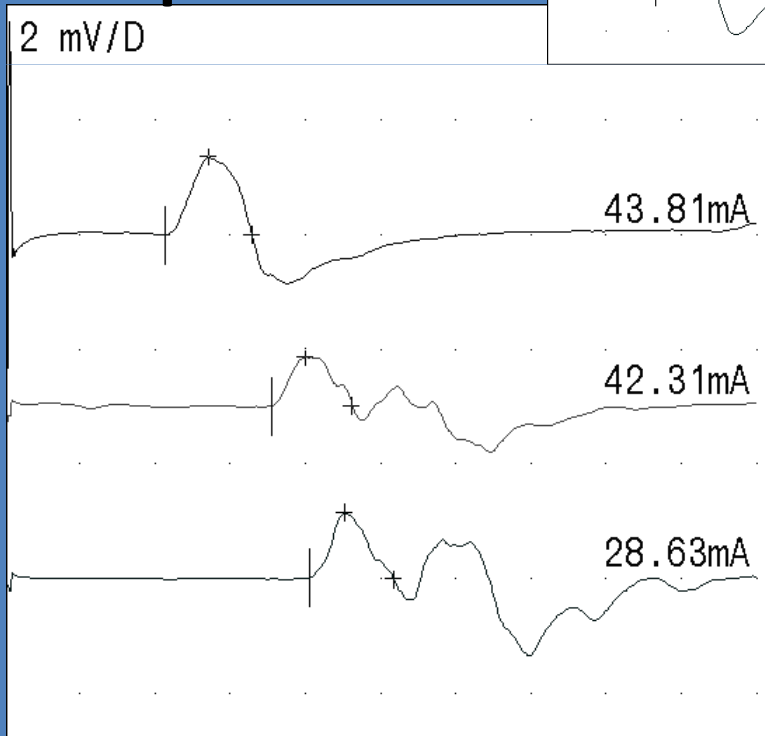
Temporális diszperzió



Konduktív blokk

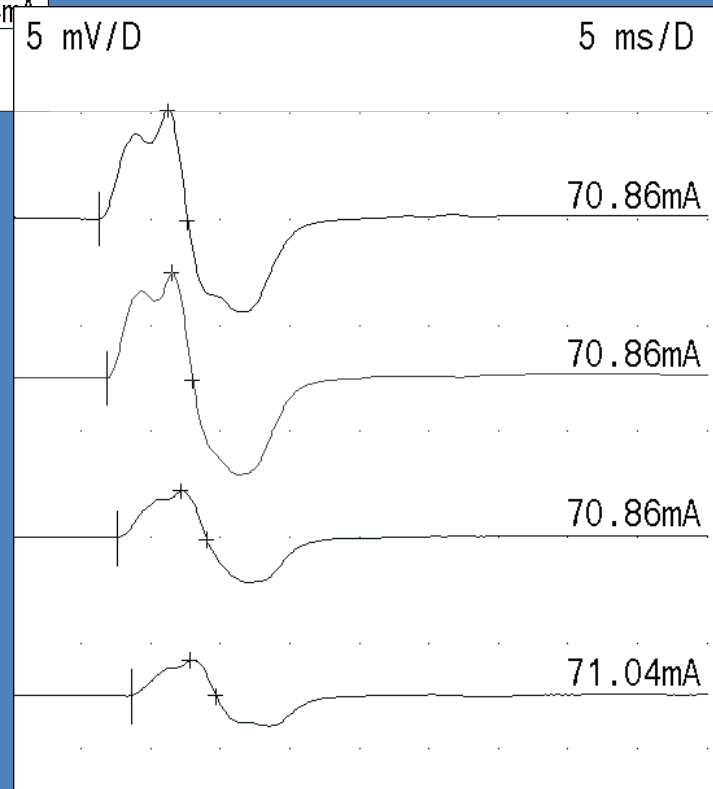


Temporális diszperzió



Normál

Konduktíós blokk



Köszönöm a figyelmet

