Retrospective analysis of repetitive nerve stimulation criteria in myasthenia gravis

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OBJECTIVE : The aim of this study was to review the parameters of repetitive nerve stimulation (RNS) used in our department for the diagnosis of myasthenia gravis (MG).

METHODS : This retrospective study included, over a period of 6 years, 127 patients with clinical suspicion of MG referred for RNS. The single fiber test was performed only in the absence of significant decrements. Anti-RACH and anti-MuSK antibodies were measured in all patients.
The study conditions for collecting decrements were as follows:

- **a minimum of four nerve/muscle pairs were tested: *nasalis*/facial, *trapezius*/spinal, *anconeus*/radial, *abductor digiti minimi* /ulnar**
- **stimulation was supramaximal and intensity remained constant throughout RNS**
- **RNS was performed at 3 Hz, except for the *trapezius*/spinal (2 Hz)**
- **the negative peak amplitude and area of compound muscle action potential (CMAP) were measured**
- **a reduction in amplitude or area greater than 10% of the 4th CMAP compared to the 1st was considered pathological. A decrease of more than 15% was considered pathological for the upper *trapezius* muscle**
- **all the tests were performed by the same operator**

Patients were divided into 3 groups (table 1):

- **D + (n=27): MG with at least two pathological decrements**
- **D - (n=19): MG with increased jitter only**
- **No-MG (n=81): no pathological decrement, no increase in jitter and no anti-RACh or MuSK antibodies**

Table 1. Cohorts of the study

Groups		Decrements	Jitter	Anti-RACh or anti-MusK
MG (n=46)	D + (n=27)	+	/	+
	D – (n=19)	-	+	+
No-MG (n=81)		-	-	-

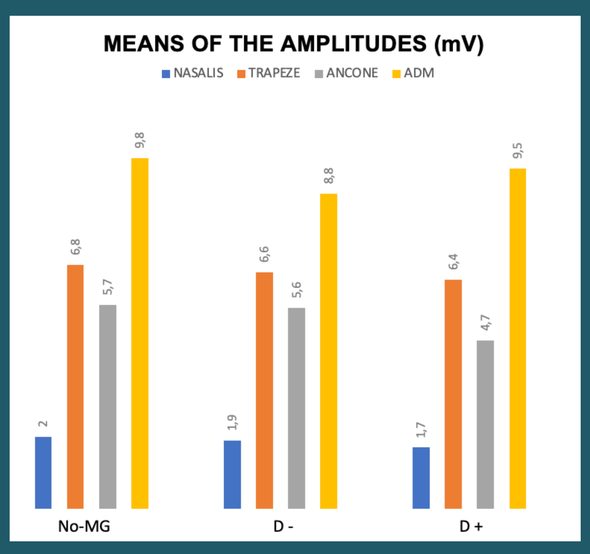
Table 2. Means, standard deviations and P95 of decrements between the 1st and 4th decrements (D1-4) and between the 1st and 2nd decrements (D1-2).

No-MG (n = 81)	*Nasalis*		*Trapezius*		*Anconeus*		*Abductor digiti minimi*	
	Mean (SD)	P95	Mean (SD)	P95	Mean (SD)	P95	Mean (SD)	P95
D 1-4	0,5 (2,1)	4	0,3 (2,2)	4	0,5 (2,8)	5	-0,1 (2,1)	3
D 1-2	0,2 (1,6)	3	-1,4 (2,7)	3	0,1 (2,4)	4	0,3 (1,7)	3

Table 3. Diagnostic sensitivity of RNS and FU with upper limit of normal (ULN) for trapezius/spinal decrement at 15% and 10%.

	RNS		SF
	ULN *trapezius* decrement		
	15%	10%	
Generalized form (n=23)	91% (21/23)	**96% (22/23)**	83% (5/6)
Ocular form (n=23)	26% (6/23)	**39% (9/23)**	84% (16/19)

RESULTS AND DISCUSSION : The mean age of the 46 patients with MG was 57 y.o. with 18 women (10 ≤ 50 y.o. & 8 > 50 y.o.) and 28 men (5 ≤ 50 y.o. & 23 > 50 y.o.).
The mean amplitudes and areas of the 1st CMAP after RNS were not statistically different between the three groups, ($p < 0.05$) (figure 1). In the No-MG group, for the 4 muscles studied, the 95th percentiles (P95) of the amplitude decrement between the 1st and the fourth CMAP (D1-4) was ≤ 5% and the P95 between the 1st and the 2nd CMAP (D1-2) was ≤ 4 % (table 2). In the D + group, the mean decrements D1-4 was greater than 10%. The mean D1-2 (7.6%) was greater than mean D2-3 (6.1%), which was itself greater than mean D3-4 (1.5%) (figure 2).
Off the 46 patients with MG, 23 had a generalized form (GF) (100% anti-RACh) and 23 had an ocular form (OF) (67% anti-RACh, 9% anti-MuSK, 24% double seronegative). RNS allowed the diagnosis of GF in 21 subjects, among the 23 identified in the study (21/23 = 91%). Five out of six patients with GF had a pathological FU exploration (5/6 = 83%). By setting a limit of normality at 10% for the *trapezius*/spinal couple, instead of 15%, the sensitivity of the RNS reaches 96% in the GF (study of the 4 muscles) (table 3).



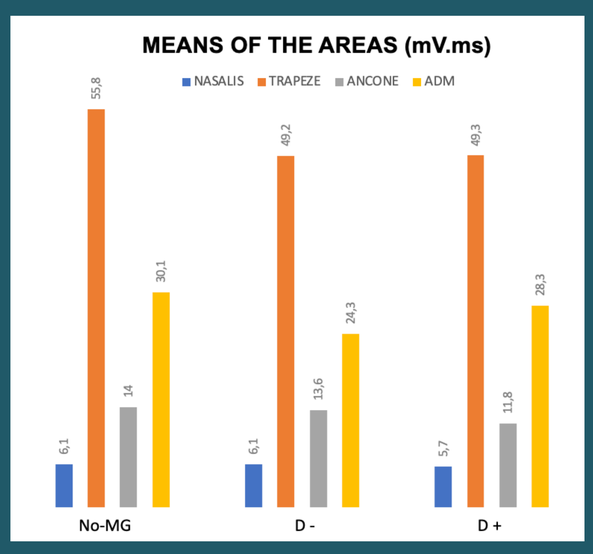


Figure 1. Mean amplitudes and areas of the 1st CMAP after RNS for the nasalis, trapezius, anconeus and abductor digiti minimi (ADM) muscles in the 3 cohorts.

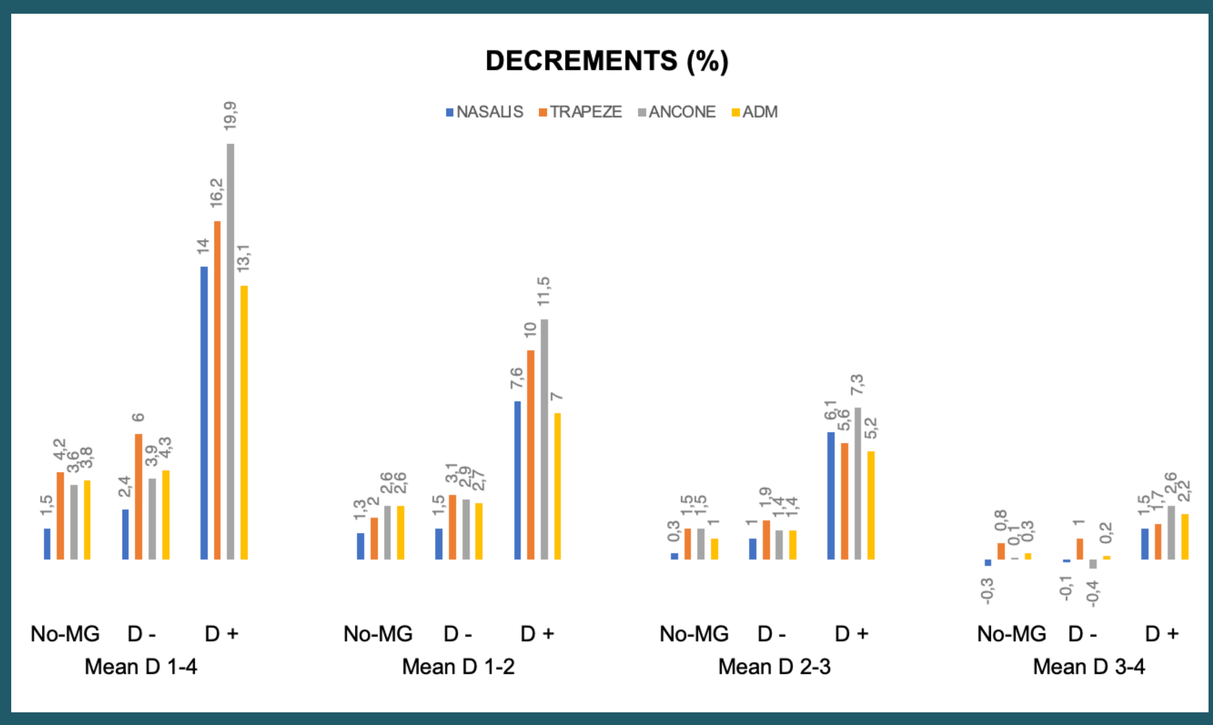


Figure 2. Mean area decrements in the 3 groups between 1st and 4th (D1-4), 1st and 2nd (D1-2), 2nd and 3rd (D2-3), and 3rd and 4th (D3-4).

CONCLUSION : A decrement less than 5% is definitively normal. A decrement between 5 and 10% is doubtful and requires completing RNS or completing it with a SF test, especially if D1-2 > 4%. Indeed, the greatest decrement between two successive CMAPs concerns D1-2. The upper limit of normal for the *trapezius*/spinal pair decrement can be reduced to 10%. With this approach, the sensitivity of RNS reaches 96% for GF.

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