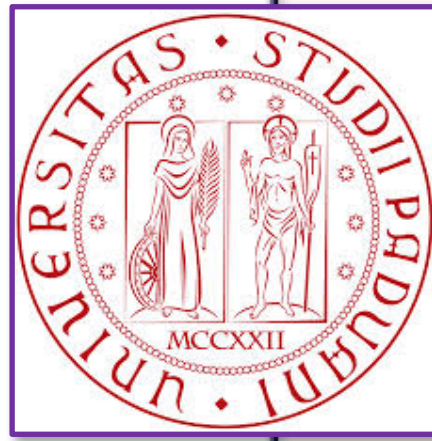# Preliminary design and test of the "6K scale" for Bulbar Symptoms evaluation in patients with SBMA.

Querin G[1], Battel I[2], Mometto L[1], Martinelli I[1], Meo G[1], Ricciardi C[1], Bertolin C[1], Pegoraro E[1], Sorarù G[1]

[1]Departement of Neurosciences, University of Padova, Padova, Italy.
[2]UOC Neurologia, Ospedale dell'Angelo, ULLS12 Veneziana, Mestre (VE), Italy

Objective

Spinal and bulbar muscular atrophy (SBMA) is a rare, late onset, X-linked neuromuscular disease characterized by slowly progressive limbs and bulbar muscle weakness and atrophy. Bulbar symptoms are a main characteristic of the disease and they may be severe and invalidating even though slow progressing. Nevertheless, a dedicated tool for their evaluation is still not available.

Aim of this study was to design and test a new scale for evaluation of bulbar function in SBMA.

Methods

- 60 genetically confirmed SBMA patients were considered.
- **Construction of the 6K-scale** → the scale is projected to evaluate the function of V, VII, IX, X, XI and XII cranial nerves (CN) and of ansa cervicalis. It is made by by 6 sub-scores and a total score (MAX score = 61).
- Patients underwent medical history collection and functional status evaluation through SBMAFRS, 6MWT, AMAT and fVC% evaluation.
- Each patient was administered the 6K-scale by two different and independent expert speech therapist.
- 18 patients underwent a re-test evaluation after 3 weeks.

V- Trigeminal nerve	*Jaw open to resistance*	Opening and strength are normal	3
		Opening is normal but strength is weak	2
		Opening is minimal and strength is absent	1
		Absence of jaw opening	0
	Jaw lateralisation	Lateralisation on both sides is normal	3
		Deficit of one side lateralisation	2
		Minimal lateralisation attempts	1
		Absence of movement	0
	Tactile sensitivity of the face (Eyebrows, Cheeks, Superior Lips, Inferior Lips)	Normal	3
		Mild	2
		Moderate	1
		Severe	0
	Tactile sensitivity of 2/3 anterior portion of the tongue (Right and Left)	Normal (6/6)	3
		Mild (5/6-4/6)	2
		Moderate (3/6-2/6)	1
		Severe (1/6-0/6)	0
VII- Facial nerve	*Eye Closure*	The eye closure is possible in both eyes	3
		One or both eyes remain slightly open (less than ½ of eye-lid)	2
		One or both eyes remain slightly open (more than 1/2 of eye-lid)	1
		Absence of eye closure	0
	Wrinkle eyebrows	Contraction is normal and symmetric	3
		Slight asymmetric contraction	2
		Severe asymmetric contraction	1
		Absence of contraction	0
	Smile	Contraction is normal and symmetric	3
		Slight asymmetric contraction	2
		Severe asymmetric contraction or slightly movements of the angles of the mouth	1
		Absence of contraction	0
	Kiss	Contraction is normal and symmetric	3
		Slight asymmetric contraction	2
		Severe asymmetric contraction	1
		Absence of contraction	0
IX- Glossopharyngeal nerve	*Elevation of the soft palate*	Contraction is normal and verbal production is not nasal	3
		Slight contraction and/or nasal quality of speech	2
		Absence of contraction and/or hypernasality.	1
	Tactile sensitivity of 1/3 posterior portion of the tongue (Right adf Left)	Tactile sensitivity is present on both sides of the tongue	3
		Tactile sensitivity is present only on one side and/or it is reduced	2
		Absence of tactile sensitivity	1
	Gag Reflex	Present	3
		Hypoactive/ Hyperactive	2
		Absence of reflex	1
X- Vagus nerve	*Voluntary Cough*	2 consecutive sound cough events	3
		1 consecutive sound cough event	2
		1 consecutive sound cough event and weak	1
		Absence of cough	0
	Vocal quality	No dysphonia	3
		Slight dysphonia	2
		Severe dysphonia	

XII- Hypoglossal nerve	*Lingual Protrusion*	The subject protrudes 2/3 of the tongue out	3
		The subject protrudes 1/3 of the tongue out and/or there is slight lateralization	2
		The subject protrudes less than 1/3 of the tongue and/or there is great lateralization	1
		Absence of movement	0
	Lingual Lateralization	Lateralization is possible without difficulties for both sides of the mouth	3
		Lateralization is possible only for one side of mouth and it is accomplished by using jaw movements	2
		The movement is limited and for one or both dental arch	1
		Absence of movement	0
	Sliding tongue	The movement is possible for the superior and inferior dental arch	3
		The movement is imprecise for one or both dental arch	2
		The movement is limited and for one or both dental arch	1
		Absence of movement	0
	Click of tongue	The click tongue is possible with sound	3
		The click tongue is weak without the specific sound	2
		There is a hint of the click of the tongue	1
		Absence of movement	0
Ansa cervicalis- XI-C1-C2	*Head control a) Up-down b) Rotation right-left c) Inclination right-left*	The three movements (a;b;c) are possible without problems	3
		The 2/3 of the movements (a;b;c) are possible	2
		One movement is possible and with difficulties controlling the head	1
		No head control	0

Scale validation

- Mean 6K score was 47 +/-5 (maximum possible score = 60).
- **Cronbach's alfa coefficient** was used to test internal consistency reliability of the scale. It was **0.85** for the total scale and respectively **0.83 for V, IX and X CN subscale and 0.84 for VII and XII CN and for ansa cervicalis subscale**, demonstrating high internal consistency.
- Inter-rater and test-retest reliability were over 0.70 as was perfect agreement test.
- Factor analysis retained a six factors solution accounting for 71.7% of the variance.
- We found strong correlation between the 6K scale total score and SBMAFRS total score and sub-scores (p<0.05). 6K scale total score correlated also with general functional scores (AMAT, 6MWT, p<0.05) as described in the table, suggesting that it could be a good integrative tool in global evaluation of SBMA patients.

	Age (ys)	Disease duration (ys)	Weight (Kg)	6MWT (meters)	AMAT score	SBMAFRS total score	SBMAFRS bulbar subscore	SBMAFRS upper limbs subscore	SBMAFRS trunk subscore	SBMAFRS lower limbs subscore	SBMAFRS respiratory subscore	fVC%
Total CN scale	r = -0.41 p = 0.01	r = -0.20 p = 0.22	r = 0.06 p = 0.68	r = 0.36 p = 0.02	r = 0.48 p = 0.01	r = 0.58 p = 0.00	r = 0.53 p = 0.02	r = 0.26 p = 0.12	r = 0.46 p = 0.02	r = 0.40 p = 0.01	r = 0.04 p = 0.70	r = 0.34 p = 0.02
V CN subscale	r = 0.09 p = 0.56	r = 0-13 p = 0.44	r = -0.41 p = 0.01	r = 0.20 p = 021	r = 0.16 p = 0.32	r = 0.24 p = 0.14	r = 0.18 p = 0.27	r = 0.23 p = 0.16	r = 0.23 p = 16	r = 0.15 p = 0.37	r = 0.02 p = 0.63	r = 0.11 p = 0.53
VII CN subscale	r = 0.34 p = 0.03	r = -0.03 p = 0.85	r = 0.20 p = 0.21	r = 0.34 p = 0.03	r = 0.40 p = 0.01	r = 0.49 p = 0.00	r = 0.36 p = 0.02	r = 0.22 p = 0.17	r = 0.47 p = 0.00	r = 0.32 p = 0.03	r = 0.10 p = 0.42	r = 0.17 p = 0.32
IX CN subscale	r = -0.17 p = 0.29	r = -0.04 p = 0.79	r = -0.09 p = 0.58	r = 0.19 p = 0.25	r = 0.33 p = 0.03	r = 0.32 p = 0.03	r = 0.27 p = 0.10	r = -0.03 p = 0.85	r = 0.36 p = 0.02	r = 0.12 p = 0.44	r = 0.12 p = 0.37	r = 0.43 p = 0.00
X CN subscale	r = -0.35 p = 0.02	r = 0.10 p = 0.54	r = -0.06 p = 0.67	r = 0.16 p = 0.32	r = 0.29 p = 0.07	r = 0.38 p = 0.01	r = 0.33 p = 0.03	r = 0.23 p = 0.16	r = 0.25 p = 0.12	r = 0.26 p = 0.10	r = 0.15 p = 0.30	r = 0.16 p = 0.33
XII CN subscale	r = -0.22 p = 0.17	r = -0.20 p = 0.07	r = 0.01 p = 0.92	r = 0.24 p = 0.14	r = 0.31 p = 0.04	r = 0.36 p = 0.02	r = 0.39 p = 0.01	r = 0.12 p = 0.45	r = 0.24 p = 0.14	r = 0.12 p = 0.10	r = 0.04 p = 0.81	r = 0.25 p = 0.14
Ansa cervicalis subscale	r = -0.14 p = 0.38	r = 0.02 p = 0.85	r = 0.01 p = 0.92	r = -0.03 p = 0.85	r = 0.03 p = 0.83	r = 0.36 p = 0.02	r = 0.09 p = 0.56	r = -0.11 p = 0.49	r = 0.13 p = 0.44	r = 0.06 p = 0.68	r = 0.12 p = 0.45	r = 0.17 p = 0.32

This is the first scale dedicated to evaluation of bulbar symptoms in SBMA. The scale has the characteristics of simple design, easy use, good reliability and validity, and good correlation between test results and global status of the patients. All these features indicate that the K6 scale can quantitatively evaluate bulbar involvement in SBMA and that is thus suitable for clinical studies.