

Orazio Pardeo¹, Marisa Maniaci¹, Natalia Longoni¹, Roberto Materia¹, Elena La Rosa¹, Rita Lauro¹, Ibrahim Said¹, Cosimo Allegra¹, Maria Sframeli¹, Eugenio Mercuri^{2,3}, Sonia Messina¹

¹Department of Clinical and Experimental Medicine, University of Messina, Messina, Italy

²Centro Clinico Nemo, Fondazione Policlinico Universitario A. Gemelli IRCCS, Rome, Italy

³Child Neurology and Psychiatry, Catholic University of Rome, Italy

Introduction

Respiratory and bulbar involvement are part of spinal muscular atrophy (SMA) clinical spectrum. There are no standardized and routinely used assessments of bulbar function in SMA. Recently the “Oral and Swallowing Abilities Tool” (OrSAT) has been developed to specifically record feeding, oral abilities and swallowing in young SMA1 patients¹.

Patients and methods

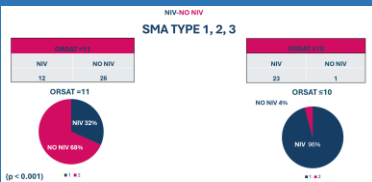
We administered the OrSAT, specifically adapted for older patients and with a score ranging from 0 to 12 and collected data on non-invasive ventilation (NIV) requirement and functional involvement of 62 SMA patients (mean age: 25,1years, SD:17,1) followed at our Centre.

Score*	0-5 m		6-9 m		10-24 m	
	1	0	1	0	1	0
1. Able to swallow thin liquids	Y	N	Y	N	Y	N
2. Able to swallow semi liquids			Y	N	Y	N
3. Able to swallow semisolids			Y	N	Y	N
4. Able to swallow solids					Y	N

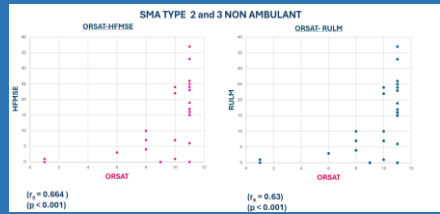
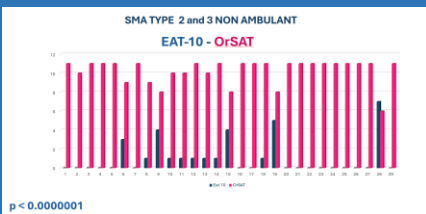
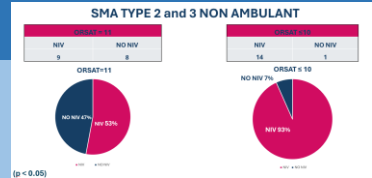
Score	1	0
5. Need for intervention	No need for intervention	Need for intervention (thickening food <input type="checkbox"/> positioning <input type="checkbox"/>)
6. Cough/signs of stagnation during meal	No Cough/signs of stagnation during meal	Cough/signs of stagnation during meal
7. Able to swallow without tiring	Able to swallow without tiring	Able to swallow but easily tired, needs to rest periodically during a meal
8. Able to complete a meal	Able to complete a meal	Unable to complete a meal
9. Duration of main meal (<45 min for semisolids and 25 min for breastfeeding)	Longer	
10. Need for Suctioning during meal time	No Need for Suctioning during meal time	Need for Suctioning during meal time
11. Able to speak one or more syllables (if age > 6 months)	Yes	No
12. Able to speak correctly one or more words (if age > 12 months)	Yes	No
		TOTAL SCORE

Results

This module showed a good responsiveness in detecting bulbar involvement across SMA types (SMA1: Mean Score: 7, SD: 4,9, 13 pts; SMA2: MS: 10, SD: 2,8, 23 pts; SMA3: MS: 12, SD: 1, 26 pts) and a strong inverse correlation with the need of nocturnal NIV ($r_s = -0.59$, $p = 0.0000001$).



In a more homogeneous subgroup (SMA2/3 not ambulant), OrSAT moderately correlated with the Hammersmith Functional Motor Scale Expanded ($r_s = 0.48$, $p = 0.006$) and strongly with the Revised Upper Limb Module ($r_s = 0.54$, $p = 0.0013$).



Conclusions

The OrSAT proved to be a valuable tool for monitoring dysphagia, showing good sensitivity in detecting bulbar involvement across different SMA types. In the subgroup SMA type 2 and 3 non ambulant we showed for the first time a strong relation between bulbar and functional involvement, more evident with the scale focusing on the upper limbs (Revised Upper Limb Module) as expected in non ambulant patients. Moreover the correlation with the respiratory function was less strong than in the whole cohort including SMA1 ($r_s = -0.39979$, $p = 0.023$). Bulbar involvement seems to develop subsequent to respiratory dysfunction, given that 55% of patients on NIV did not present it, whereas all patients with OrSAT impairment required NIV support. We propose the modified OrSAT as a valuable tool to improve the study and management of the complex interactions between respiratory and bulbar function in advanced disease stage.

References

1. Berti B, et al. *J Neuromuscul Dis* 2021;8:589–601