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Trajectory of change in the swallowing status in spinal muscular atrophy type I

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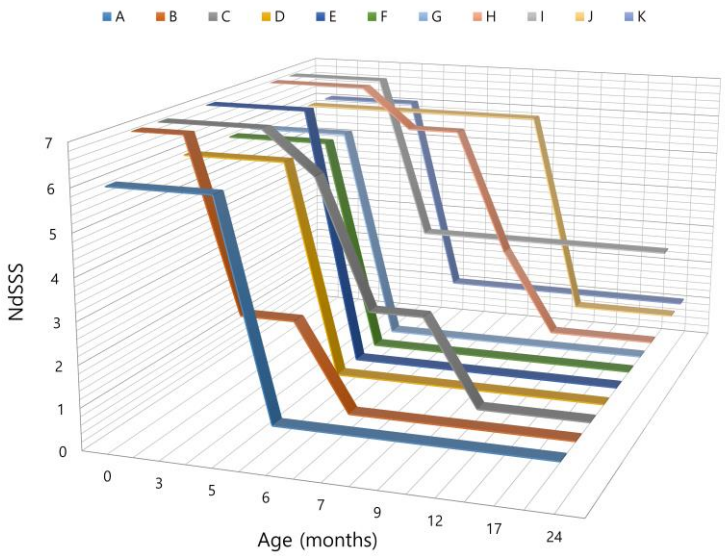
Aim
Spinal muscular atrophy (SMA), an autosomal recessive genetic disorder, is characterized by progressive muscle weakness and atrophy. An objective description of swallowing difficulty in patients with SMA type I is currently lacking making the management of patients with such conditions rather challenging. This study aimed to describe the change in progressive swallowing dysfunction in 11 subjects with SMA type I from birth to 2 years of age using the Neuromuscular Disease Swallowing Status Scale (NdSSS) and videofluoroscopic swallowing study (VFSS).

Methods
Retrospective chart reviews were performed. The NdSSS was used to describe the actual swallowing situation in patients with SMA type I and VFSS was used to assess swallowing function in an objective manner.

Results
Upon analysis, we found that the swallowing function generally deteriorated in patients with SMA type I at approximately 6 months of age, and the average age at which tube feeding was initiated was approximately 6.8 ± 2.0 months. However, there was wide variation in the period when the main route of feeding was changed from totally oral to tube feeding (from 5 months to 12 months). Five subjects with SMA type I had VFSS data. In some cases, the evidence of laryngeal aspiration was obtained via the VFSS at very early stages of the disease. Conversely, there were some cases in which mainly oral feeding was maintained up to 12 months and evidence of aspiration was not observed in the VFSS.

Conclusion
An individualized approach is essential, as the timeline of deterioration in swallowing function varies widely in patients with spinal muscular atrophy type I.



Trajectory graph of NdSSS in patients with type I SMA according to age.