

Dysphagia in children with spinal muscular atrophy type II

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Introduction (1)

Spinal muscular atrophy (SMA) (1 : 6000 newborns) [1]

Degeneration of anterior horn cells (motor neurons) in the spinal cord

Weakness and muscular atrophy

Four types of SMA

SMA type II is characterized by the ability to maintain an unsupported sitting position.

[1] Lefebvre S, Burglen L, Reboullet S, Clermont O, Burlet P, Viollet L et al. Identification and characterization of a spinal muscular atrophy-determining gene. Cell 1995; 80(1): 155-65.

Introduction (2)

In patients with SMA type II:

- problems with mouth opening, chewing, swallowing (more with solid food) and choking more than in patients with SMA type III and IV [2]
- (Aspiration) pneumonias: a video fluoroscopic swallow study is recommended [3]

[2] Groot IJMd, de Witte LP. Physical complaints in ageing persons with spinal muscular atrophy. J Rehabil Med 2005; 37(4): 258-62.

[3] Wang CH, Finkel RS, Bertini ES, Schroth M, Simonds A, Wong B et al. Consensus statement for standard of care in spinal muscular atrophy. J Child Neurol 2007; 22(8): 1027-49.

Introduction (3)

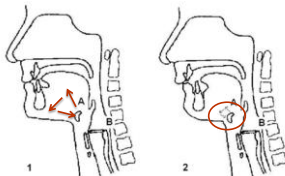
Hypothesis about swallowing problems:

1. weak movements of the tongue and a restricted opening of the mandible [4];
2. weak movements of the submental muscle group (SMG) leading to dysfunctional protection of the airway and opening of the upper esophageal sphincter (UES);

[4] Houston K, Buschang PH, Iannaccone ST, Seale NS. Craniofacial morphology of spinal muscular atrophy. Pediatr Res 1994; 36(2): 265-9.

Introduction (4)

3. lumbar lordosis associated with anterior tilted pelvis and problems with head balance due to weak neck musculature causing compensatory behavior like a retracted neck [5].



[5] Engel-Hoek Lvd, de Swart BJ, Erasmus CE, Groot IJMd. Is Head Balance a Major Determinant for Swallowing Problems in Patients With Spinal Muscular Atrophy Type 2? J Child Neurol 2008; 23(8):19-21.

Aim of the study

To determinate the underlying mechanisms of the dysphagia in SMA type II:

primary neurological problems or

biomechanical problems (compensatory posture components) or

a combination

Methods (1)**Patients and controls:**

Children with SMA type II attending the multidisciplinary outpatient clinic,
with complaints about either mouth opening, chewing or swallowing

Matched healthy controls

The study was approved by the Committee on Research Involving Human Subjects of Arnhem and Nijmegen in the Netherlands.

Methods (2)

Patient characteristics, general and physical abilities (Motor Function Measure scale) [6a]

Feeding and swallowing assessment:

The Nijmegen Dysarthria Scale (NDS) [6b]

Self composed questionnaire for complaints about swallowing and duration of meals (five point scale)

The Dysphagia Disorder Survey (DDS) [7]

[6a] Barard C, Payan C, Hodgkinson I, Fermanian J. A motor function measure for neuromuscular diseases. Construction and validation study. *Neuromuscul Disord*;15:463-470.
[6b] Kruuji S, Kalf H. Dysarthriëonderzoek. *Logopedie en Foniatrie* 2007;12:412-417.
[7] Sheppard JJ. Dysphagia Disorder Survey and Dysphagia Management Staging scale (Manual, Dutch translation by Visuelgers, R. en Penning, C.). 1 ed. Rotterdam: Faculteit Geneeskunde en Gezondheidswetenschappen. 2002.

Methods (3)

Assessment of mandibular function

Mandibular function impairment questionnaire [8]

Mouth opening

[8] Stegenga, B., de Bont, L. G., de, L. R., & Boering, G. (1993). Assessment of mandibular function impairment associated with temporomandibular joint osteoarthritis and internal derangement. *J.Orofac.Pain*, 7, 183-195.

Methods (4)**Registration of swallowing:**

Surface electromyography (sEMG) of the submental muscle group (SMG)

Video fluoroscopic swallow study (VFSS), only in the patient group

5 ml thin liquid with a syringe placed on the tongue

5 g solid food (pureed potato) with a spoon [9]

Two different postures:

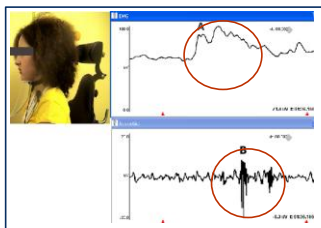
retracted posture (RP) and forward posture (FP)

Control group: normal position (FP) and in a position with a retracted neck and mandible (RP)

[9] Ozdemirkiran T, Seclil Y, Tarlaci S, Ertekin C. An EMG screening method (dysphagia limit) for evaluation of neurogenic dysphagia in childhood above 5 years old. *Int J Pediatr Otorhinolaryngol* 2007; 71(3): 403-7.

Methods (5)

sEMG in combination with cervical auscultation and video



The sEMG was used to evaluate the duration (in seconds) and muscle activity (mean amplitude in μV) of each swallow act [10].

[10] Cray MA, Baldwin BO. Surface electromyographic characteristics of swallowing in dysphagia secondary to brainstem stroke. *Dysphagia* 1997; 12(4): 182-7.

Methods (6)

The video files of VFSS were captured and stored on the Digital Swallowing Workstation (Kay Pentax)

A three point scale (0, normal; 1, slightly disturbed; 2, disturbed) was used to quantify different parameters [11,12].

The video files were reviewed by a second SLT and after discussion consensus was achieved about the scores for the parameters [13].



[11] Higo R, Nito T, Tayama N. Videofluoroscopic assessment of swallowing function in patients with myasthenia gravis. *J Neurol Sci* 2005; 231(1-2): 45-8.
[12] Rogers B, Arvedson J, Buck G, Smart P, Msall M. Characteristics of dysphagia in children with cerebral palsy. *Dysphagia* 1994; 9(1): 69-73.
[13] Scott A, Perry A, Bench J. A study of interrater reliability when using videofluoroscopy as an assessment of swallowing. *Dysphagia* 1998; 13(4): 223-7.

Statistical analysis:

Descriptive statistics (mean and standard deviation) for the sEMG signal and the duration of swallowing per group.

Descriptive statistics in terms of median for the scores on the VFSS.

The difference between the amplitude of the sEMG signal in the different postures and the duration of swallowing was tested using the nonparametric Wilcoxon test. The test was two-tailed and conducted at the 5% significance level.

Results

6 patients 6.05 -13.04 of age (9.7 ± 2.9);

6 matched healthy controls 6.04 - 13.08 of age (9.6 ± 2.8)

In all patients:

- sitting position with retracted mandible
- tongue movement problems: elevating their tongue tip and clacking their tongue was abnormal
- fasciculations and atrophy of the tongue

Results and discussion

Only six patients, with some variability in functional ability and complaints about feeding and swallowing

They all have a sitting position with the head more or less in retraction

They show the same patterns during the registration of swallowing:

abnormal patterns during swallowing solid food with piecemeal deglutition, vallecular residue and residue above the UES

measured with sEMG of the SMG with a significant relation to head position.

Discussion

Relationship between feeding and swallowing problems and recurrent pneumonias

Consensus Statement for Standard of Care in Spinal Muscular Atrophy: a VFSS should always be performed if there are concerns about the safety of swallowing [3]

Presence of recurrent pneumonia: a potential indicator of aspiration (silent or indirect)

Post swallow residue: risk for aspiration when the airway reopens [11]

[3] Wang CH, Finkel RS, Bertini ES, Schroth M, Simonds A, Wong B et al. Consensus statement for standard of care in spinal muscular atrophy. *J Child Neurol* 2007; 22(8): 1027-49

[11] Arvedson JC. Pediatric videofluoroscopic swallow studies: A professional manual with caregiver guidelines. San Antonio: Communication Skill Builders, Physiological Group, 1998.

Conclusion

SMA II patients have less problems with swallowing liquid than solid food and seem to benefit from a more forward position when swallowing solid food.

Conclusion:

The underlying components of the swallowing problems in children with SMA type II:

neurological component (tongue and efficiency of SMG)

biomechanical component (compensatory posture).

An integrated treatment with:

an adapted posture during meals

adjusting meals (avoiding or reduce solid food)

advice of drinking water after meals clearing the oropharyngeal area to prevent aspiration pneumonias.