Dysphagia in children with spinal muscular atrophy type II

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Introduction

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.

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]

Introduction (2)

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);

Introduction (3)

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]

Methods (3)

Assessment of mandibular function

Mandibular function impairment questionnaire [8]

Mouth opening

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)

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].

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].

References:
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]

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.