

SHORT REPORT

Ptosis aggravates dysphagia in oculopharyngeal muscular dystrophy

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Background: Ptosis and dysphagia are important features in oculopharyngeal muscular dystrophy (OPMD).

Objective: Retroflexion of the head is a well known compensatory mechanism for ptosis, but generally retroflexion has a negative effect on swallowing. We hypothesised that severity of ptosis is related to degree of retroflexion and that this compensation is responsible for deteriorating dysphagia.

Methods: Nine OPMD patients were examined in the conditions “head position adapted to ptosis” and “head position slightly flexed”. Ptosis was quantified by photogrammetry and retroflexion of the head by digital photographs. The severity of dysphagia was measured using visual analogue scales (VAS) and by calculating swallowing volumes and oropharyngeal swallow efficiency (OPSE) based on videofluoroscopy.

Results: Statistical analyses show a significant relationship between ptosis and degree of retroflexion. The degree of retroflexion of the head correlated significantly with VAS scores and with the maximum swallowing volume. The slightly flexed head position significantly improved VAS scores as well as swallowing volumes and OPSE.

Conclusion: In OPMD patients, ptosis significantly correlates with retroflexion of the head, which has a negative effect on swallowing. Subjective and objective reduction of swallowing problems was found when patients were instructed to eat and drink with a slightly flexed head position.

Oculopharyngeal muscular dystrophy (OPMD) is an adult onset, usually autosomal dominant, slowly progressive muscle disorder.¹ The most striking feature is bilateral ptosis, which ultimately limits the vision. To compensate for ptosis, patients contract their frontal muscles and hold their head in retroflexion (astrologist's view).¹ Another important feature is dysphagia, which can cause life threatening complications, for instance aspiration pneumonia.

We hypothesised that in OPMD patients, the combination of astrologist's view and pre-existing dysphagia can aggravate the dysphagia. In healthy people, retroflexion of the head has a negative effect on swallowing by widening the laryngeal entrance, narrowing the valleculae, and decreasing the upper oesophageal sphincter relaxation, causing less adequate bolus passage.^{2–3}

In this study, we tested the hypothesis that the severity of ptosis is related to the degree of retroflexion of the head and that this is responsible for a deterioration in dysphagia in OPMD patients. In addition, we investigated the effect of a slightly flexed head position on dysphagia.

METHODS

Subjects

Nine consecutive, genetically proven OPMD patients (five women and four men) were included in the study. Mean age was 57 years (range 43–69 years). Patients who had undergone a cricopharyngomyotomy or were going to undergo operation were excluded, as were those who had had ptosis corrected surgically. Informed consent was obtained from each patient and the local ethical committee approved the study.

Procedure

The ophthalmologist quantified the ptosis by photogrammetry. Frontal digital photographs of each patient's face were taken while the patient was positioned in a standard ophthalmic headrest. The patients were instructed to relax their face and to look at a fixed distant point straight ahead. A 245 mm lens was used and the camera to subject distance was 3 m. A metal ruler was fitted on the headrest in the plane of the patient's eyes to serve as a scale. Ptosis was defined as the distance between the edge of the upper eyelid and the midpoint of the pupil in millimetres.⁴ Ptosis of both eyes was measured and the average ptosis was calculated.

During the study, each patient was examined in two different conditions: first in the condition “head position adapted to ptosis”, that is patients were asked to eat and drink without any instructions, and second in the condition “head position slightly flexed”, that is after instructions. In the condition “head position adapted to ptosis” retroflexion was measured using digital photography.

Functional swallowing measurements were carried out by a speech therapist. Patients were asked to quantify their disabilities as regards eating and drinking for the conditions described above on a visual analogue scale (VAS). To objectively investigate the dysphagia, the maximum swallowing volume (the amount of water the patient is able to take with only one swallow) was measured in both conditions. The oropharyngeal swallow efficiency (OPSE) was also calculated in both conditions using videofluoroscopy with 20 ml of thin liquid barium and 20 ml of thick liquid barium. OPSE is a measure to quantify the patient's ability to transport a food bolus efficiently and safely from the oral cavity to the oesophagus.⁵ It is the ratio between the amount of the bolus that is swallowed during the first swallow and the oropharyngeal transit time. OPSE increases when a larger amount of material is swallowed during the first swallow and when the oropharyngeal transit time decreases.

Statistical analyses

Statistical analyses were conducted with SPSS 11.0 (SPSS, Chicago, IL, USA). The Spearman correlation coefficient was

Abbreviations: OPMD, oculopharyngeal muscular dystrophy; OPSE, oropharyngeal swallow efficiency; VAS, visual analogue scale

Table 1 Characteristics of and results in OPMD patients

Patient characteristics			Ptosis (mm)	Head retro-flexion	VAS scores, fluid		VAS scores, solid		Swallow, volumes (ml)		OPSE (20 ml thin liquid)		OPSE (20 ml thick liquid)	
No.	Age	Sex	Average	Degrees	Retr	Flex	Retr	Flex	Retr	Flex	Retr	Flex	Retr	Flex
1	55	M	1.18	24	78	84	56	65	25	30	41.85	60.80	50.53	52.36
2	56	M	3.46	1	96	97	86	96	35	40	64.00	81.07	47.30	64.00
3	62	F	2.54	16	100	100	71	71	—*	—*	57.60	60.63	40.96	43.52
4	63	M	0.61	7	83	90	56	73	12	20	90.00	132.35	54.05	75.89
5	43	F	2.84	1	96	96	77	87	45	55	64.39	79.17	40.76	68.97
6	60	M	0.53	26	72	76	47	70	10	17	43.60	75.89	70.83	73.28
7	57	F	2.66	4	85	91	56	66	30	38	—*	—*	—*	—*
8	52	F	1.76	24	83	87	53	81	5	8	60.34	89.29	—†	—†
9	69	F	2.45	19	84	92	82	90	15	30	—‡	—‡	—‡	—‡

*Patients 3 and 7 were excluded since they became emotional and anxious because of the risk of aspiration; †in patient 8, residue after swallowing in the extended position could not be rinsed away which made interpretation of the videofluoroscopy unreliable; ‡patient 9 aspirated during the first examination with thin liquid barium and had to be excluded from further radiological examination.

retr, retroflexion: "head position adapted to ptosis"; flex, flexion: "head position slightly flexed".

used for calculating the correlation between the degrees of retroflexion of the head and the severity of the ptosis, between the degrees of retroflexion and the VAS scores in the condition "head position adapted to ptosis", and between the degrees of retroflexion and the maximum swallowing volume. Using the Wilcoxon non-parametric test, the effect of changing head position from retroflexion to slight flexion on the VAS scores, the maximum swallowing volume, and the OPSE were tested. The level of significance was set at 0.05.

RESULTS

In the condition "head position adapted to ptosis", the degrees of retroflexion of the head (table 1) correlated significantly with the average ptosis ($r = -0.824$; $p = 0.006$) as well as with the VAS scores in the same condition (eating: $r = 0.701$; $p = 0.035$; drinking: $r = 0.767$; $p = 0.016$), and also

with the maximum swallowing volumes ($r = -0.831$; $p = 0.011$).

In the condition "head position slightly flexed", the VAS scores improved significantly compared to "head position adapted to ptosis" (eating: $z = -2.533$, $p = 0.011$; drinking: $z = -2.375$, $p = 0.018$). The swallowing volumes were significantly larger in the "head position slightly flexed" than in the condition "head position adapted to ptosis" ($z = -2.527$; $p = 0.012$) (fig 1). This corresponds with the significant increase in the OPSE in the condition "head position slightly flexed" (thin liquid: $z = -2.240$; $p = 0.025$; thick liquid: $z = -2.201$; $p = 0.028$).

DISCUSSION AND CONCLUSION

This study shows that retroflexion of the head because of ptosis results in deterioration of pre-existing dysphagia in OPMD patients. In addition, we showed that a slightly flexed head position improves swallowing. Changing head position also improves the subjective VAS scores as well as the objective swallowing volumes and radiological results. The slightly flexed position reduces the time needed for eating and drinking, reduces the amount of residue, and thus reduces the life threatening risk of aspiration in OPMD.

It is remarkable that patients are unaware of the negative effect of the retroflexion of the head on dysphagia. They accept swallowing problems as part of pharyngeal muscle weakness in OPMD. Furthermore, even instructed patients report that they often forget to change head position during eating because eating is a social activity during which looking around and facing others sitting at the table is normal behaviour, while looking continuously downwards, to improve swallowing, is not common. Additional support for the correlation between ptosis and dysphagia is a patient's experience of improvement of swallowing after surgical ptosis correction; this patient was unaware of the effect of head position on swallowing.

Based on our data, speech therapists, neurologists, and ophthalmologists can alleviate swallowing problems in OPMD by teaching patients to eliminate compensatory retroflexion of the head during eating and drinking.

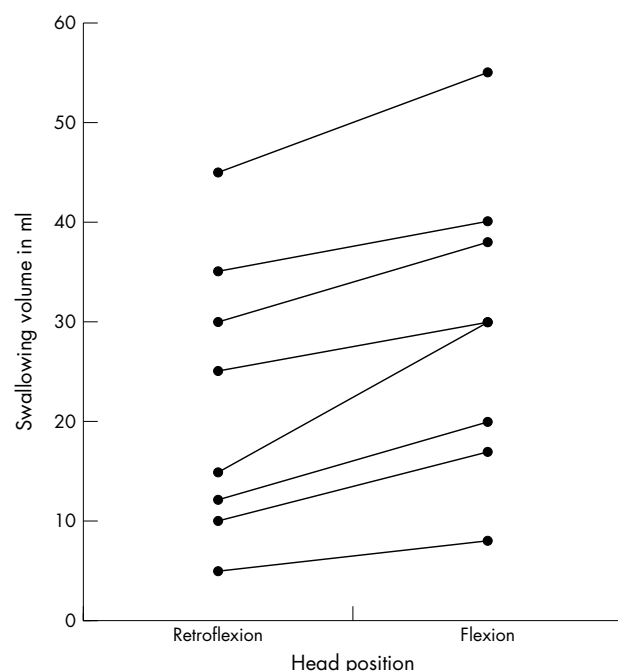


Figure 1 Maximum swallowing volumes per patient under the conditions retroflexion ("head position adapted to ptosis") and flexion ("head position slightly flexed").

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