Oropharyngeal swallowing in craniocervical dystonia

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PAPER

Objective: To clarify the pathophysiology of dysphagia by electrophysiological methods.

Method: Electrophysiological methods related to oropharyngeal swallowing were used to investigate 25 patients with cervical dystonia and 25 age matched normal volunteers.

Results: Dysphagia was suspected in 36% of patients with cervical dystonia on the basis of clinical assessment. The incidence of dysphagia increased to 72% on electrophysiological evaluation of pharyngeal swallowing. Submental muscle electromyographic (EMG) and laryngeal relocation times were significantly prolonged and the triggering time to swallowing reflex was significantly delayed. Some abnormalities seen in cricopharyngeal sphincter muscle EMG indicated that the striated sphincter muscle is hyperreflexive in some patients.

Conclusion: Neurogenic dysphagia was more prominent and longer lasting than mechanical dysphagia, which was transient and varied from patient to patient. Although these electrophysiological methods were not suitable for detecting anatomical changes during swallowing, as in videofluoroscopic studies, observations supported the neurogenic cause of dysphagia in patients with any kind of cervical dystonia.

METHOD

Patients

Twenty-five patients with cervical dystonia (13 male and 12 female) were investigated. The average duration of illness ranged from a few months to many years. Their average age was 42.8 years (range 19–68). Patients were divided into three groups according to their clinical presentations:

- Pure spasmodic torticollis group: 11 patients with leftward pulling in 3, rightward pulling in 6, and anterocollis in 2.
- Oromandibular and laryngeal dystonia group: 7 patients with spasmodic dysphonia in 3, oromandibular dystonia in 3, and lingual dystonia in 1. One patient with oromandibular dystonia also had anterolaterocollis but the others had no torticollis at all.
- Generalised dystonia plus spasmodic torticollis: 7 patients.

Method

Oropharyngeal function was clinically evaluated in each patient according to our simple grading of dysphagia.14 This classification is based purely on symptoms and signs of difficulty swallowing. The degree of dysphagia was graded as follows:

- grade 1: no clinical signs or symptoms of dysphagia were present.
- grade 2: very mild dysphagia was suspected in clinical examination but the patient never complained directly of dysphagia.
- grade 3: the patient complained of dysphagia and this was supported by other clinical signs.
- grade 4: obvious clinical signs and symptoms of dysphagia including aspiration; dysphagia severe enough to necessitate non-oral feeding.

The first group of patients was investigated either just before the first injection of botulinum A toxin or over the two months after the last botulinum A toxin injection. The remaining patients were not given any chemodenervation therapy or surgical intervention.

Electrophysiological evaluation of oropharyngeal swallowing

EMG methods to investigate the nature of oropharyngeal swallowing have been previously published.15 16 Firstly, the onset and duration of oropharyngeal swallowing were recorded from the submental or suprahyoid muscles (SM-EMG). SM-EMG surface electrodes were usually placed at the midline bilaterally under the chin.

Secondly, the vertical movements of the larynx during swallowing were monitored by a piezoelectric sensor placed between the thyroid and the cricoid cartilages. This supplied...
Dysphagia in cervical dystonia

A Normal subject

Laryngeal sensor signal

B Torticollis

Figure 1: Laryngeal sensor signals (upper traces in A and B) and integrated submental electromyographic (SM-EMG) activities (lower traces in A and B) obtained from a normal subject (A) and a patient with torticollis (B). Points A and C denote the onset and end of SM-EMG activity. Points 0 and 2 denote the onsets of laryngeal upward and downward movement, respectively. The oblique arrow from A to 0 [A-0 interval] denotes the time interval from the onset of voluntarily triggered swallowing to the onset of reflex swallowing as shown by the onset of laryngeal upward movement. Extreme prolongation of the A-C interval and a delay in triggering time of the swallowing reflex (oblique arrow) are observed in the patient (B). Amplitude calibration is 30 and 50 μV, respectively, for SM-EMG traces. Time calibration is 200 ms all traces. All traces are averages of five responses.

The individual results obtained from the 25 patients with cervical dystonia were compared with the upper limits of deglution parameters obtained from 25 normal age matched subjects. The abnormal prolongation of A-C interval and a delay in triggering time of the swallowing reflex were observed in 17 patients, mostly with generalised dystonia plus oromandibular dystonia groups and only two patients had grade 2 dysphagia. On the other hand, five of seven patients with generalised dystonia plus cervical dystonia had trouble swallowing and their degree of dysphagia changed from grade 2 to grade 3. Thus, the proportion with dysphagia was 36% among patients with cervical dystonia based on clinical assessment. The dysphagia was mild and some patients compensated for their swallowing problems by special head and neck manoeuvres while swallowing. The majority of patients did not complain of any kind of deglutition problems. Despite this lack of complaints, the electrophysiological evaluations showed that “subclinical dysphagia” is not uncommon and precedes medical or surgical treatment.

Dysphagia limit

Figure 2 illustrates the dysphagia limits of one patient with and one without clinical dysphagia. Non-dysphagic patients swallowed up to 20 ml of water in a single attempt, whereas patients with dysphagia divided the bolus into two or more parts while trying to swallow 20 ml or less of water. A dysphagia limit of < 20 ml of water was found not only in dysphagic patients but also in others with normal swallowing (dysphagia grade 1). The abnormal dysphagia limit was encountered in 18 patients, making the frequency of dysphagia 72% in patients with cervical dystonia. Thus, subclinical dysphagia was determined in 36% of all patients with cervical dystonia by using these electrophysiological methods. The dysphagia limit was often 15–20 ml water in patients with cervical dystonia with subclinical dysphagia, whereas patients with clinical dysphagia were not able to swallow 1–10 ml in one attempt. The dysphagia limit was also useful to understand the effect of neck movements on swallowing. Some patients prevent the effect of sudden neck movements by fixing the head or changing the position to anteflexion during swallowing. In such patients, having the head and neck held by the examiner improved their dysphagia limit, as fig 3 illustrates. Unfortunately, the effect of holding the head could not be investigated in all patients because of difficulty in obtaining physical cooperation.

Single bolus analysis

Table 1 shows individual results of the single bolus analysis of swallowing 3 ml of water. The most frequent and suggestive finding was the significant prolongation of SM muscle complex activity (A-C interval in table 1 and fig 1). The age dependant increase in the duration of SM muscle activity in the patients was accounted for by comparison with three age groups of normal subjects (17–39, 40–59, and 60–81 years, published previously). The abnormal prolongation of SM muscle complex activity during 3 ml water swallowing was found in 17 patients, mostly with generalised dystonia plus torticollis. This may mean that 68% of patients with cervical dystonia have abnormally high and long firing of the motor

RESULTS

Clinical assessment

In the clinical assessment, 9 of 25 patients with cervical dystonia had expressed some subjective complaints or had objective clinical signs suggesting oropharyngeal dysphagia. Swallowing problems were mild in the spasmodic torticollis and the oromandibular and laryngeal dystonia groups and only two patients had grade 2 dysphagia. On the other hand, five of seven patients with generalised dystonia plus cervical dystonia had trouble swallowing and their degree of dysphagia changed from grade 2 to grade 3. Thus, the proportion with dysphagia was 36% among patients with cervical dystonia based on clinical assessment. The dysphagia was mild and some patients compensated for their swallowing problems by special head and neck manoeuvres while swallowing. The majority of patients did not complain of any kind of deglutition problems. Despite this lack of complaints, the electrophysiological evaluations showed that “subclinical dysphagia” is not uncommon and precedes medical or surgical treatment.

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data about the timing of laryngeal elevation and suspension during the pharyngeal phase of the reflex response. The total duration of SM-EMG activity was determined from the A-C interval and the laryngeal relocation time was measured from the onsets of the upward and downward signals of the laryngeal sensor, or the 0–2 time interval, during swallowing (fig 1).

The triggering time of the pharyngeal phase or reflex swallowing was calculated from the time interval between the onsets of SM-EMG and the first deflection of the laryngeal sensor movement (the A-0 interval in fig 1).

Thirdly, using the same EMG methods described above, it was possible to diagnose objectively the existence of dysphagia in suspected cases by an indicator we called the dysphagia limit. While 3 ml of water was swallowed in single bolus analysis, the dysphagia limit was determined by increasing the amount of water swallowed step by step from 3 to 20 ml of water. The analysis time was taken as 10 seconds for this purpose instead of the 2 or 5 seconds allowed for single bolus analysis. Normal subjects can swallow a 20 ml bolus of water in a single attempt. Dysphagic patients are unable to swallow 20 ml water volumes at one go and divide the bolus into two or more parts within 8 seconds. All patients were investigated by this method in a sitting position with or without their head being held by one of the examiners.

Fourthly, in nine patients, EMG activity of the cricopharyngeal (CP) muscle of the upper oesophageal sphincter was recorded through a concentric needle electrode inserted percutaneously and directed postero-medially in the neck.

The individual results obtained from the 25 patients with cervical dystonia were compared with the upper limits of deglutition parameters obtained from 25 normal adult subjects divided into the three age groups of 17–39 years, 40–59 years, and 60–81 years. The individual values of CP sphincter EMG during swallowing were also compared with the normal values obtained from 25 normal age matched adult subjects with a mean age of 42.2 years (range 17–69, 13 male, 12 female). Informed consent was obtained from all patients and normal subjects. The investigation was approved by the ethical committee of our university hospital.
units of the submental laryngeal elevator muscles. The results were also significant (mean (SEM) 1200.3 (63.3) ms v 857.9 (23.2) ms, p < 0.0001). The second common abnormality was prolongation of the laryngeal relocation time during swallowing (0–2 interval in table 1 and fig 1). This finding was encountered in 10 of the 25 patients (42%). This time interval is intimately related with the time required for the pharyngeal phase of the swallowing reflex. This result was also significant (674.1 (26.2) ms v 571.5 (14.2) ms, p < 0.0001). Lastly, the triggering time of the swallowing reflex was delayed in eight patients (A-0 interval in table 1 and fig 1). This was also significant in all patient groups (410.5 (48.1) ms v 261.4 (20.9) ms, p < 0.0001). Overall, 31% of patients exhibited this delay but the incidence was higher in patients with oromandibular and laryngeal dystonia (four of seven patients). Thus, one or more of these neurogenic signs of swallowing disorder were found in 19 patients (76%) by single bolus analysis.
CP sphincter EMG

The CP sphincter muscle of the pharyngoesophageal segment could be investigated in only nine patients. Among these patients CP-EMG during swallowing was found to be normal in two patients who had a “CP-EMG pause” lasting 400–600 ms and with forebursts and rebound bursts of CP tonic activity (fig 4). However, the remaining seven patients (one with oromandibular and laryngeal dystonia, one with spasmodic torticollis, and the rest with generalised dystonia) were found to have some abnormalities indicating a kind of hyperreflexion of the sphincter muscle. Among these, the unexpected burst of motor units within the CP-EMG pause was common (fig 4). Other less frequent abnormalities encountered were shortening of the pause and premature closure of the CP sphincter EMG. It was noted that the absence of the foreburst was also quite frequent in patients with cervical dystonia but not usually observed in hyperreflexive CP-EMG. It was noted that the absence of the CP sphincter EMG. It was noted that the absence of the foreburst was also quite frequent in patients with cervical dystonia but not usually observed in hyperreflexive CP sphincter.

Relation between swallowing and sternocleidomastoid muscle activity

In four patients with spasmodic torticollis, the SM-EMG and the hyperreflexive sternocleidomastoid (SCM) EMG (needle recording) were recorded synchronously. There was no swallowing activation as traced by SM-EMG and laryngeal sensor during the SCM burst activity due to clinical spasms. Similarly, the swallowing movement did not produce any EMG activation in the SCM muscle. During the dysphagia limit investigation with boluses from 3 to 20 ml of water, in all four cases the second swallow was not synchronous with SCM activation. Some patients had discovered special head or neck positions and holding the head in a stable position sometimes improved swallowing (fig 3). Mechanical dysphagia seemed to be a transient phenomenon. In other patients with clinical dysphagia, holding the head or neck fast or maintaining a lateral spine resting position did not prevent repetitive swallowing or improve the dysphagia limit, though these procedures were not performed in all patients.

Summing up, involuntary neck movements did not continuously produce dysphagic signs clinically or electrophysiologically; nor did swallowing aggravate the head or neck positions in our patient groups.

DISCUSSION

Dysphagia was suspected in 36% of patients with cervical dystonia on the basis of clinical assessment. The incidence of dysphagia increased to 72% on electrophysiological evaluation of oropharyngeal swallowing. Clinical and videofluoroscopic evaluations have also indicated a high incidence of swallowing disorders in patients with cervical dystonia before any treatment such as botulinum toxin injection or rhizotomy. The incidence of dysphagia varied between 22 and 100% of patients (mostly over 50%). The incidence of dysphagia increased more significantly after botulinum toxin injection or rhizotomy. Similarly, dysphagia in patients with spasmodic dysphonia has been reported before and after treatment of this condition.

In our patient groups 11 had pure spasmodic torticollis and 7 had torticollis with generalised dystonia. However, in our 7 patients with oromandibular and laryngeal dystonia there were no abnormal neck movements except in one patient. Despite the heterogeneity of the patients with cervical dystonia, the clinical and electrophysiological findings were similar for each group.

The major finding of this study was that the neurogenic nature of dysphagia was more prominent and longer lasting than the mechanical dysphagia, which was transient and varied from patient to patient. Although our electrophysiological
methods were not suitable for detecting anatomical changes during swallowing, as in videofluoroscopic studies, the following observations supported the neurogenic cause of dysphagia in patients with any kind of cervical dystonia:

Firstly, similar clinical and electrophysiological findings were found in three groups of patients including the oromandibular and laryngeal dystonia group, who did not have spasmodic torticollis. Therefore, head or neck movement is not essential for dysphagia.

Secondly, some patients maintained a certain position to compensate for the adverse effect of neck movement during swallowing. Holding or fixing the head was beneficial in some other patients (fig 3). In clinically affected patients and some other patients, stabilising the head and neck was not effective in changing the patients' dysphagia clinically, nor were the dysphagia limits improved.

Thirdly, no EMG relation was observed between swallowing and EMG activity of the SCM muscle. Videofluoroscopic evaluation of patients with cervical dystonia generally has shown delayed initiation of swallowing and pharyngeal residue. These abnormalities are regarded as neurogenic dysfunction in oropharyngeal assessment with videofluoroscopy. In our patients the most frequently encountered electrophysiological abnormality was prolongation of SM muscle complex activity during swallowing (68% of all patients). Though prolongation of the laryngeal relocation time and triggering of the swallowing reflex were less frequent than SM muscle activity, they were seen in 42% and 31%, respectively, of all patients with cervical dystonia. Such findings are observed mostly in patients with Parkinson's disease. They are not related to mechanical dysphagia. In normal subjects, when the head is rotated towards any side or held in retroflexion, the dysphagia limit may change but the single bolus parameters of the SM-EMG complex and sensor recording never change.

Fourthly, electrophysiological abnormalities were frequent and all were pertinent to neurogenic dysphagia. Videofluoroscopic evaluation of patients with cervical dystonia generally has shown delayed initiation of swallowing and pharyngeal residue. These abnormalities are regarded as neurogenic dysfunction in oropharyngeal assessment with videofluoroscopy. In our patients the most frequently encountered electrophysiological abnormality was prolongation of SM muscle complex activity during swallowing (68% of all patients). Though prolongation of the laryngeal relocation time and triggering of the swallowing reflex were less frequent than SM muscle activity, they were seen in 42% and 31%, respectively, of all patients with cervical dystonia. Such findings are observed mostly in patients with Parkinson's disease. They are not related to mechanical dysphagia. In normal subjects, when the head is rotated towards any side or held in retroflexion, the dysphagia limit may change but the single bolus parameters of the SM-EMG complex and sensor recording never change.

Fifthly, CP sphincter EMG during swallowing was not normal in patients with cervical dystonia, especially in those in the generalised dystonia group. Some of the CP-EMG findings were compatible with the hyperreflexive/hypertonic nature of the upper oesophageal sphincter observed in suprabulbar involvement of the bulbary swallowing centre in the central nervous system.

Lastly, aggravation of dysphagia following medical and surgical treatment of spasmodic torticollis points to the neurogenic nature of dysphagia; otherwise, an improved head or neck position would have eased the mechanical dysphagia. Botulinum toxin treatment and rhizotomies may increase dysphagic signs or unmask the potential inherent neurogenic nature of dysphagia in these patients. However, transient brisk neck spasms may still produce a transient and sporadic negative effect on swallowing at any time.

It is not known how and where the neurogenic dysphagia of cervical dystonia is located in the central nervous system. This awaits further studies in line with the pathophysiology of the general nature of dystonias.

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Figure 4 Superimposed laryngeal sensor signals and cricopharyngeal (CP) EMG signals recorded from a patient with spasmodic dysphonia with dysphagia (A) and a normal subject (B) during wet swallowing. In the patient with spasmodic dysphonia, the unexpected burst of motor unit action potentials (MUAP) within the swallowing pause of the CP muscle is clearly seen (arrow). The CP-EMG pause is shorter in the patient with spasmodic dysphagia than in the normal subject. The third trace is obtained from conventional EMG recordings. The second and fourth EMG traces are rectified and integrated. The first trace shows the laryngeal sensor movements during swallowing by the patient. Amplitude calibration is 50 µV for CP-EMG traces. Time calibration is 200 ms in all traces.
**REFERENCES**


