Dysphagia in Patients With Frontotemporal Lobar Dementia

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Background: Hyperorality, compulsive eating and aspiration because of food gorging, has been described in patients with frontotemporal lobar dementia (FTLD), but swallowing function in this population has not been reported.

Objective: To identify the swallowing status in a sample of patients with FTLD.

Design: Case series.

Setting: Referral center, ambulatory care.

Patients: A consecutive series of referred patients with 3 variants of FTLD were asked to participate. Twenty-one patients were enrolled, including 9 with frontotemporal dementia, 7 with progressive nonfluent aphasia, and 5 with semantic dementia.

Intervention: The patients underwent a fiberoptic endoscopic examination of swallowing to assess their ability to swallow liquids and food.

Main Outcome Measures: Presence of dysphagia and nature of impaired swallowing.

Results: Of the 21 patients, 4 caretakers reported swallowing difficulties. An instrumental examination revealed moderate swallowing abnormalities in 12 of the 21 patients. These abnormalities were not explained by compulsive eating behaviors, but seemed to reflect deficits in cortical and subcortical pathways connecting with the brainstem swallowing center.

Conclusions: When assessed via instrumentation, swallowing abnormalities are found in many patients with FTLD. The appearance of dysphagia signals progression of FTLD to brainstem systems.

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AMONG PATIENTS WITH frontotemporal lobar dementia (FTLD), there is considerable diversity in symptoms.1 Clinical subtypes present initially with behavioral features (frontotemporal dementia [FTD]) or language impairment (semantic dementia [SD] or progressive nonfluent aphasia [PNFA]). Although most patients eventually develop abnormal clinical neurological signs, including parkinsonism or motor neuron disease, a few present with early neurological deficits. Sometimes, these deficits remain as secondary features, while other times, they portend an evolving problem that eventually is diagnosed as amyotrophic lateral sclerosis (ALS), progressive supranuclear palsy (PSP), or cortical basal ganglionic degeneration (CBD).

A recent retrospective study2 analyzed early predictors of mortality in patients with FTLD via retrospective review of records of 96 patients up to the time of FTLD diagnosis. Patients with deficits that preceded diagnosis, such as mutism or dysphagia, had a shorter survival time, while those with preassessment behavioral abnormalities had a longer survival. The researchers postulated that the early appearance of neurological deficits, semimutism or mutism, or dysphagia indicated a more rapidly progressive disorder, involving more extensive degeneration of cortical and subcortical structures. All patients with dysphagia later developed ALS, suggesting that the early swallowing impairment was due to the as-yet undiagnosed ALS. Thus, early detection of swallowing problems in patients with FTLD had diagnostic and prognostic implications.

Pneumonia is a common cause of death in patients with FTLD.3 In a recent study2 of 73 deceased patients with FTLD, the most common causes of death were other diseases of the central nervous system (n=37) and pneumonia (n=18). Asphyxia accounted for death in 3 patients. The pneumonia was likely an aspiration pneumonia due to underlying dysphagia,
but the study did not probe further for this association. Pneumonia is also a common cause of death in patients with PSP and CBD,\(^4,5\) and patients with ALS, PSP, or CBD are known to have swallowing problems. However, dysphagia is not well documented in those with FTLD in the absence ALS, PSP, or CBD.

There are 2 reports describing swallowing function in those with FTLD without concomitant ALS, PSP, or CBD. One described a patient with PNFA who manifested an “apraxia of swallowing.”\(^6\) The problem, visualized fluoroscopically, was difficulty initiating the swallow, with food and liquid remaining in the patient’s mouth for long periods. Otherwise, the swallow was unremarkable.

Another study\(^7\) investigated eating and swallowing behaviors in those with FTLD (only FTD and SD) and Alzheimer disease by administering a questionnaire to the relatives of 91 patients. While most of the items (30/36) probed for changes in appetite, food preference, eating habits, and other oral behaviors, 6 asked about swallowing problems. The severity of dementia was similar in the 2 patient groups. The frequency of reported problems was higher in the FTLD group than in the Alzheimer disease group, especially for the eating domains. Overall, dysphagia symptoms were the least reported problem in all patient groups, occurring in only 20% to 30% of patients. Patients with Alzheimer disease reportedly developed swallowing symptoms earlier in the course of the disease than patients with FTD and SD. Their method for assessing dysphagia only used a questionnaire, and swallowing was not formally assessed.

In summary, the prevalence of swallowing problems in patients with FTLD is unknown. While compulsive, rapid, careless eating can lead to airway obstruction and asphyxia, it does not generally cause aspiration pneumonia, which is a common cause of death in this population. If FTLD leads to swallowing deficits, regular monitoring and management of this behavior may prevent complications. Moreover, if dysphagia is a sign of early ALS, PSP, or CBD, this would yield valuable prognostic information for patients, their caregivers, and the health professional, and would enable them to optimize management.

This study formally assesses swallowing function in patients with FTD, PNFA, and SD without known ALS, PSP, or CBD to determine whether dysphagia was present and, if so, to characterize the nature of the problem.

**PATIENTS**

Thirty consecutive patients diagnosed as having FTD, PNFA, or SD were referred for evaluation of swallowing function from the Memory and Aging Center at University of California, San Francisco. Consent was obtained with a research protocol that included patient and family consent, and was approved by the institutional review board. In all, there were 15 subjects with FTD, 8 with PNFA, and 7 with SD. None of these patients had PSP, CBD, or ALS as a second diagnosis. A control group of 9 individuals similar in age to the patients but with no neurological disease and normal swallowing symptoms underwent the same examinations.

**PROCEDURES**

Each patient, and caretaker when present, was interviewed about the patient’s eating and swallowing behaviors. Each patient was given a formal examination to assess tongue, lip, laryngeal, and pharyngeal symmetry, force, and control of movement. Specific tasks probed for oral apraxia. Speech was judged for the presence of dysarthria or apraxia of speech. The results of these examinations will be reported separately.

A fiberoptic endoscopic examination of swallowing (FEES) was conducted to formally evaluate the oropharyngeal stage of swallowing.\(^8\) The patient took food and liquid to eat and drink while the hypopharyngeal and laryngeal structures were kept in view and the bolus was visualized. Chewable food, pureed food, and liquid were ingested. While one examiner (S.E.L.) passed the endoscope, another observed the patient’s eating habits. The examination was videorecorded and reviewed by 3 judges (S.E.L. and 2 others) for the variables of interest. An ordinal scoring scale was developed, including 58 variables. Variables to score included such items as adequacy of mastication, leakage of food into the pharynx during mastication, location of the bolus head at swallow onset, amount and location of residue after the swallow, presence and time of aspiration, response of the patient to bolus presence, dysphagia severity, and deterioration during the study. Half of the examinations were scored by 2 judges (S.E.L. and 1 other) independently, and interrater reliability was computed. Disagreements were discussed until consensus was reached.

**DATA ANALYSIS**

All results were tallied, organized on a spreadsheet, and analyzed. Because of the few patients in each subgroup of FTLD, all were pooled into 1 group when compared with the healthy controls for statistical purposes. The data were a mix of ordinal and quantitative measures; therefore, nonparametric statistics were used (Mann-Whitney test for 2 independent groups).

**RESULTS**

Of the 29 patients referred for evaluation, 5 refused the FEES procedure (3 in the FTD and 2 in the SD group) and 3 recorded swallow studies were lost to technical error, leaving 21 patients with complete results. The final group included 9 patients with FTD, 7 with PNFA, and 5 with SD. The mean age of the patients was 61.4 years (standard deviation, 6.1 years), the mean time since onset of FTLD was 5.6 years (standard deviation, 1.7 years), and the mean Mini-Mental State Examination score was 22.2 (of a possible 30) (standard deviation, 6.2). None of these subject characteristics was noticeably different among the 3 subgroups. A healthy control group of 9 subjects who underwent a FEES was comparable in age (mean, 60 years).

The interjudge reliability for all examinations scored by 2 examiners (S.E.L. and/or 2 others) was good, with 85% of the examinations showing identical ratings and 100% being only 1 point apart on any single rating.

**REPORTED EATING HABITS**

All the patients with FTD, most of the patients with SD, and most of the patients with PNFA were reported to have altered their eating patterns. Interestingly, the caretak-
ERS were far more likely to report problems than the patients, who, for the most part, had no complaints. The patients in the 3 subgroups had similar profiles, with a strong tendency to eat rapidly and compulsively (eating everything in front of them without stopping) and to take large bolus sizes. Food rituals were not reported for the PNFA group but were common in the SD and FTD groups (e.g., needs to eat chips with every meal or positions food in a certain order on the plate). Patients in all 3 subgroups had gained weight in the past 6 months. None of the patients reported any difficulty swallowing, but 4 caretakers described a problem. The most common type of problem described was one of occasional choking episodes. Table 1 summarizes the subject characteristics and reported eating patterns.

ENDOSCOPIC EXAMINATION OF SWALLOWING

The performance of our 9 healthy control subjects on an FEES was used as reference data representing normal swallow function. Their scores agreed for the most part with the literature on normal swallow measures, except for the fact that one of our healthy subjects aspirated once. The performance of our subject groups was compared with these normative data, using scores more than 1 standard deviation away from the mean to indicate a possible deviancy and scores more than 2 standard deviations away to represent “abnormal” function.

Although some of the patients with FTLD displayed unusual behaviors, the reported compulsive, rapid, overeating habits were only apparent in a few of the patients during the FEES procedures. This took the form of continual eating and drinking without pause until the food was gone. The presence of the endoscope and the clinical environment may have inhibited some of these eating behaviors in the patients and may have altered the results of the study.

Table 2 summarizes the results of the FEES procedures in the different subject groups. Two clearly aberrant swallow behaviors emerged in the FTD groups. First, when the subjects were masticating food, before swallowing it, the subjects with FTLD showed a clear tendency to let food leak into the pharynx for an excessive time. Our healthy control subjects let food leak for a mean of 1.20 seconds during mastication before they initiated the swallow. Occasionally, a bolus leaked for a longer time, up to 9 seconds once (mean longest leakage time, 2 seconds; standard deviation, 3 seconds). However, all the FTLD groups showed longer leakage times. The mean leakage time for the FTD group was 3.06 seconds, and the longest mean leakage time was 15.78 seconds. One subject let the bolus leak into the pharynx for 45 seconds! During this period, the subjects seemed oblivious to the fact that their throats were filling up with food while they were talking and chewing.

As a consequence of the long bolus leakage time during mastication, patients let the bolus fall lower in the throat before the swallow was initiated. At the time of swallow onset, our healthy subjects either still held the food bolus in the mouth or had let it slip to the base of tongue. No healthy subject let the food bolus fall lower than the tongue base. The average bolus position at the onset of the swallow for the patients with FTLD was the pit of the valleculae, with the lowest mean spillage point midway down the pharynx. One third of the patients let the bolus fall to the pit of the piriform sinuses or the laryngeal rim before the swallow was triggered. These locations represented the furthest that a bolus can move without penetrating the laryngeal vestibule and clearly increased the risk of aspiration.

The second abnormal pattern displayed by some of our patients was incomplete bolus clearance during the swallow. Pharyngeal clearance of food was complete in every healthy control subject, even in our 83-year-old woman. However, one third of the subjects with FTLD left residue of food in the pharynx after the swallow. In most cases, the amount of residue was mild, but in 2 cases, the residue filled 1 or more of the cavities within the pharynx. Incomplete bolus clearance is a sign of reduced force of contraction of the muscle groups that propel or squeeze

<table>
<thead>
<tr>
<th>Subject Group</th>
<th>Age, y*</th>
<th>Age at Onset, y*</th>
<th>MMSE Score*</th>
<th>Reported Altered Eating Patterns</th>
<th>Reported Weight Change</th>
<th>Reported Swallow Impairment</th>
</tr>
</thead>
<tbody>
<tr>
<td>Healthy controls (n = 9)</td>
<td>60.00 (18.36)</td>
<td>NA</td>
<td>NA</td>
<td>NA</td>
<td>5 (56%) gained weight</td>
<td>0</td>
</tr>
<tr>
<td>FTD (n = 9)</td>
<td>60.10 (4.33)</td>
<td>54.42 (3.32)</td>
<td>20.25 (7.46)</td>
<td>9 (100%) of all subjects: 5 compulsive, 4 rapid, 4 large bolus, 4 food rituals, and 4 gained weight</td>
<td>5 (56%) gained weight</td>
<td>2 (22%)</td>
</tr>
<tr>
<td>PNFA (n = 7)</td>
<td>64.14 (8.65)</td>
<td>61.71 (7.50)</td>
<td>22.43 (5.97)</td>
<td>4 (57%) of all subjects: 3 compulsive, 1 rapid, and 2 gained weight</td>
<td>2 (29%) gained weight</td>
<td>2 (29%)</td>
</tr>
<tr>
<td>SD (n = 5)</td>
<td>58.40 (2.07)</td>
<td>54.40 (1.67)</td>
<td>25.00 (3.39)</td>
<td>4 (80%) of all subjects: 4 rapid, 2 large bolus, 2 compulsive, 1 food rituals, and 2 gained weight</td>
<td>2 (40%) gained weight</td>
<td>0</td>
</tr>
</tbody>
</table>

Abbreviations: FTD, frontotemporal dementia; MMSE, Mini-Mental State Examination; NA, data not applicable; PNFA, progressive nonfluent aphasia; SD, semantic dementia.

*Data are given as mean (standard deviation).
the bolus through the pharynx (tongue, pharynx, and larynx). The high incidence of this deficit was totally unexpected. The clinical examinations had detected impaired motor control in the tongue and pharynx in about half of these same patients, but the FEES procedure had revealed weakness in an additional 3 patients.

Despite their swallowing abnormalities, only 1 patient with FTLD (with SD) aspirated once during the swallowing examination. If the subjects had manifested the compulsive eating habits reported in the interviews, the incidence of aspiration may have been higher.

In summary, more than half (n=12) of the 21 patients with FTLD scored abnormally (2 standard deviations away from the normal mean) on 1 or more swallowing variables. The overall severity of the dysphagia, however, was generally categorized as mild because the patients rarely aspirated and bolus clearance was fairly good. Seventeen variables from the FEES procedure were analyzed with a Mann-Whitney nonparametric test. Results yielded a significant difference between the control subjects and the subjects with FTLD on 5 variables, all relating to the food swallows: leakage time during mastication (P=.007), swallow onset time (P=.03), typical (P=.04) and lowest (P=.005) bolus location at swallow onset, and reduced bolus clearance (P=.05).

In this study, swallowing behavior in patients with FTLD was characterized. Data from the 3 major subgroups of FTLD were inspected separately, but for most of the measures, performance did not greatly vary across groups. The patients with SD and FTD had more compulsive eating behaviors than the PNFA group, but this pattern was seen in all 3 groups. Although "hyperphagia" was not directly observed during most of the FEES procedures, the family report of this behavior helped to explain why, in some patients, mild dysphagia coupled with compulsive eating could easily lead to aspiration.

The endoscopic procedure (FEES) rather than fluoroscopy was chosen for this project so that the subjects could eat real food at their natural rate and take typical bolus sizes. Only the endoscopic examination could have captured the distinct pattern manifested by many of the patients (that of excessive food leakage into the throat during oral mastication). During fluoroscopy, the patient’s natural pattern of letting the bolus leak during mastication might not have been captured. Interestingly, the leakage tended to increase as the study progressed, after 10 or more swallows of the same food.

Healthy persons, in natural eating conditions (with no instructions when to swallow), do not always swallow food just as it leaves the mouth. A typical pattern is to “dump” food to the valleculae just as mastication ends, but before the swallow begins.10,11 This “dumping,” or transitional, time averages about 1 to 2 seconds, similar to the time shown by the healthy subjects in our study. Most patients in this study were clearly beyond the limits of healthy in terms of number of seconds of leakage. The fact that the leakage took place during mastication, when volitional control of behavior predominates and cortical and subcortical circuits are most active, suggests a breakdown in normal monitoring of these neural centers.

The other major problem that occurred in 8 (38%) of the 21 patients was incomplete bolus clearance through the pharynx. This was a surprising finding because none of the patients had known weakness of the bulbar musculature. Bolus clearance through the mouth and throat requires a fair degree of strength in the tongue and pharyngeal and laryngeal muscles. Bolus clearance is not controlled by cortical structures, but is more likely mediated by brainstem centers with facilitating input from

### Table 2. Results of the FEES Procedure

<table>
<thead>
<tr>
<th>Subject Group</th>
<th>Typical/Longest* Time</th>
<th>Typical†/Lowest‡ Location of Food at Onset of Swallow$</th>
<th>Typical/Lowest Location of Liquid at Onset of Swallow$</th>
<th>Typical/Worst§ Amount of Food Residue Left After the Swallow$</th>
<th>Aspiration Severity Rating#</th>
</tr>
</thead>
<tbody>
<tr>
<td>Healthy controls (n = 9)</td>
<td>1.20 (standard deviation, 1.32)/2.00 (standard deviation, 3.00)</td>
<td>0.33 (standard deviation, 0.50)/0.50 (standard deviation, 0.53)</td>
<td>0.33 (standard deviation, 0.71)/1.75 (standard deviation, 1.50)</td>
<td>0/0</td>
<td>0.11 (standard deviation, 0.33)</td>
</tr>
<tr>
<td>FTD (n = 9)</td>
<td>3.06/15.78</td>
<td>0.89/1.22</td>
<td>0/0.44</td>
<td>0.11/0.44</td>
<td>0</td>
</tr>
<tr>
<td>PNFA (n = 7)</td>
<td>1.29/4.57</td>
<td>0.71/1.00</td>
<td>0.14/0.29</td>
<td>0/0.57</td>
<td>0</td>
</tr>
<tr>
<td>SD (n = 5)</td>
<td>2.40/6.60</td>
<td>1.00/1.20</td>
<td>0.60/1.00</td>
<td>0.20/0.40</td>
<td>0.40 (liquid only, once)</td>
</tr>
</tbody>
</table>

**Abbreviations:** FEES, fiberoptic endoscopic examination of swallowing; FTD, frontotemporal dementia; PNFA, progressive nonfluent aphasia; SD, semantic dementia.

*Significant difference (P=.007) between healthy controls and patients with frontotemporal lobar dementia (FTLD) pooled.
†Significant difference (P=.04) between healthy controls and patients with FTLD pooled.
‡Significant difference (P=.005) between healthy controls and patients with FTLD pooled.
§There is a scale from 0 to 4 for location of bolus: 0 indicates the bolus is behind the tongue; 1, the bolus is at the base of the tongue or valleculae; 2, the bolus moves to lateral channels or the tip of the epiglottis; 3, the bolus is in piriforms or touches the laryngeal rim on the sides or back; and 4, the bolus falls into the laryngeal vestibule or is aspirated before the swallow. 
‖Significant difference (P=.05) between healthy controls and patients with FTLD pooled.
¶There is a scale from 0 to 4 for rating of aspiration: 0 indicates none; 1, trace amount liquid only; 2, regular aspiration of liquid; 3, occasional aspiration of food; and 4, regular aspiration of food.

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subcortical centers and indirect pathways. It will be important to observe the patients with reduced bolus clearance over the coming years to see if they develop other neurological diagnoses, such as ALS, PSP, or CBD.

The importance of uncovering swallow abnormalities in these patients cannot be underestimated. The fact that early dysphagia is predictive of shorter survival duration in patients with FTLD\(^1\) is more ominous when one realizes that dysphagia was documented in 12 of the 21 patients tested in this study and was found in all 3 variants. Compulsive eating behavior added to dysphagia increases the risk of aspiration and its complications. Aspiration pneumonia is a common cause of death in those with FTLD, and in those with PSP, CBD, and ALS.\(^2,3\) If neurologists are made aware of dysphagia in their patients early in the course of their disease, and the problem is managed by a speech pathologist specializing in dysphagia, there is a good chance that the complications of this problem can be postponed.

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