Swallowing Disorders 105

Adult Neurologic Disorders
Michael A. Crary

OUTLINE

Preliminary Considerations: Swallowing Symptoms and Neurologic Deficits
Brief overview of functional neuroanatomy relative to swallowing functions 41
Cortical Functions
Cortical functions and swallowing impairment 42
Swallowing Deficits in Hemispheric Stroke Syndromes
Treatment considerations 44
Swallowing Deficits in Dementia
Treatment considerations 49
Swallowing Deficits in Traumatic Brain Injury
Treatment considerations 52
Subcortical Functions
Subcortical Functions and Swallowing Impairment:
Parkinson’s Disease 53
Treatment considerations 54
Brainstem Functions
Brainstem functions and swallowing impairment 56
The role of the cerebellum in swallowing 57
Lower Motor Neuron and Muscle Disease
Lower motor neuron functions and swallowing impairment 57
Muscle diseases and swallowing impairment 59
Idiopathic or Iatrogenic Disorders of Swallowing That Resemble Neurogenic Dysphagia
Take Home Notes 61
62

OBJECTIVES
1. Explain why it is important to possess a basic understanding of the nervous system to clinically manage swallowing disorders resulting from neurologic disease.
2. Name some of the sensorimotor characteristics associated with impairments at different levels of the nervous system.
3. Identify some of the dysphagia characteristics that might be seen in diseases affecting various levels of the nervous system.
4. Describe some of the dysphagia-related problems that might be seen in patients with neurologic disease.
5. Describe some aspects of change in dysphagia over time in neurologic diseases.
6. Identify some of the more common treatment issues, decisions, options, and practices in different forms of neurogenic dysphagia.

PRELIMINARY CONSIDERATIONS: SWALLOWING SYMPTOMS AND NEUROLOGIC DEFICITS
Swallowing disorders are symptoms of underlying disease processes. One implication of this perspective is that swallowing disorders in patients with neurologic disorders should manifest the characteristics of damage to different areas of the nervous system. This premise has long been
accepted in the arena of motor speech disorders (dysarthria). For example, spastic dysarthria results from damage to the upper motor neuron system governing speech production. Upper motor neuron damage results in specific patterns of neuromotor impairment, including spasticity, slowed movement, exaggerated reflexes, and reduced range of movement. The characteristics of spastic dysarthria are believed to be the direct result of spasticity in the corticobulbar system governing speech production. Patients with spastic dysarthria demonstrate a slow rate of speech, limited movement of the speech articulators, equalized stress patterns, and other characteristics reflecting the underlying neuromotor characteristics of spastic weakness.

A similar framework helps clinical specialists evaluate and plan treatment for patients with swallowing disorders resulting from neurologic deficit. Patients with damage to upper motor neuron systems characteristically demonstrate spastic weakness with resultant slowness and reduced range of movement. This may translate to reduced speed of swallowing (i.e., a delay in initiating one or more components of the swallow) or reduced range of movement in the swallowing mechanism (i.e., reduced transport of the bolus contributing to postswallow residue). To understand better the potential clinical applications of such a framework, clinical specialists must be familiar with neuroanatomy, neurologic functions and dysfunctions of various nervous system components, and sensorimotor components of swallowing at different stages of the swallow. A summary of some common neurologic functions associated with various levels within the central nervous system follows.

**Brief Overview of Functional Neuroanatomy Relative to Swallowing Functions**

Adequate swallowing depends heavily on adequate movement of structures within the upper aerodigestive tract. Motor and sensory systems work together to produce movement, including movement associated with swallowing. However, in clinical practice motor and sensory functions are described separately as they may relate to impaired swallowing physiology. To facilitate a clinical perspective, a top-down approach to the nervous system is followed in which sensory and motor components are described at each level. Figure 3-1 is a simplified schematic depicting each “level” of the nervous system. Table 3-1 summarizes neurobehavioral and sensorimotor functions associated with each level.

**CORTICAL FUNCTIONS**

Functional control of sensorimotor behaviors in the human cortex frequently is described in reference to various areas or regions. The frontal lobe cortex is deemed responsible for multiple aspects of motor control, ranging from intent and initiation of movement to coordinating a movement in time and space to executing the movement in an organized and timely fashion. In general, parietal lobe regions are responsible for recognizing and interpreting sensory functions. These functions might include identifying the presence of a sensory stimulus or the interpretation of a sensory stimulus in reference to an appropriate motor response. Sensorimotor impairments resulting from cortical damage may vary in response to the location of neurologic deficit,
extent of the deficit (larger areas of damage are believed to result in more severe or widespread behavioral impairments), and whether the neurologic damage is unilateral or bilateral.

Other important functions housed within the cortex are those of human communication and cognition. Damage to primarily the left side of the brain may result in a number of difficulties in the ability to communicate. Focal attention is frequently afforded to the area of the inferior frontal lobe and superior temporal lobe, although damage to these areas often is accompanied by damage to adjacent motor control areas of the frontal lobe and/or sensory control areas of the parietal lobe. Cognitive deficits associated with cortical dysfunction may present in various forms with different levels of severity and different clinical courses depending on the location of damage and the nature of the underlying disease process.

Arriving to and leaving from the cortex are the major sensory and motor tracts within the central nervous system. Damage to the sensory tracts arriving in the primary sensory strip of the anterior parietal lobe results in loss of recognition of sensory stimuli in the corresponding body area. Damage to the motor tracts leaving the primary motor strip in the posterior frontal lobe (upper motor neurons) results in paresis or paralysis of the corresponding body area. Sensory or motor deficits are similar regardless of the location of the damage along the tracts. For example, cortical level damage to the upper motor neuron system results in the same type of motor weakness as subcortical or brainstem upper motor neuron damage.

**Cortical Functions and Swallowing Impairment**

If motor functions of the cortex range from intent to execution, then swallowing deficits resulting from cortical damage may range from no observable swallow activity to poorly coordinated execution of the act of swallowing. In considering these possibilities, frequent cortical pathologic conditions such as stroke, dementia, and traumatic brain injury (TBI) should be reviewed.

Before reviewing dysphagia characteristics in various cortical pathologies, a worthwhile question to ask is “Where is swallowing function represented in the human cortex?” Given the complexity of motor control involved in oropharyngeal swallowing, it is logical to implicate the frontal cortex, specifically areas involved in various components of motor control. In fact, results of both animal and human studies using lesion or cortical stimulation techniques implicate the importance of the lateral frontal cortex, the inferior frontal lobule, and the insula in various motor acts associated with feeding and swallowing. Recent studies using functional magnetic resonance imaging (fMRI) implicate a wide range of cortical, subcortical, and brainstem structures involved in swallowing performed by healthy volunteers.\(^2\)\(^4\) Not surprisingly, the primary motor and sensory cortical areas consistently participated in swallowing function. In a comparison of ischemic stroke patients with dysphagia and stroke patients without dysphagia, the internal capsule emerged as the only brain region significantly associated with dysphagia. However, other areas of the sensorimotor cortex and the basal ganglia also were frequently associated with the presence of dysphagia in stroke patients.

Although findings from many studies implicate a dysphagia based in poor motor control resulting from damage to the anterolateral and precentral frontal cortex, no consensus exists concerning the specific characteristics of these dysphagias. Still, hemispheric damage to frontal areas underpinning motor control resulting in direct movement deficits should raise significant clinical concern for the presence of dysphagia.

What about cortical or hemisphere lesions that impair sensory function? These sensory areas of the hemisphere may be important in understanding swallowing functions and impairments. In fact, some studies report that many stroke patients with dysphagia have damage to the parietal lobe with associated sensory deficit.\(^6\) Primary sensory areas of the cortex have extensive interconnections with the motor areas of the cortex. Sensory function is deemed important in the control of voluntary movement. Beyond direct sensory loss, we should consider conditions in which the patient cannot interpret sensory information, for example, neglect. Patients with neglect may not respond to a stimulus in the swallowing tract (food or liquid bolus), not because of direct sensory loss, but because of a cortical deficit in processing and interpreting sensory information. In at least one study, hemispatial neglect was related to clinician recommended nonoral intake of food and liquid, but not severity of dysphagia, in patients evaluated 3 days after hospital admission for stroke.\(^7\) Unfortunately, these investigators did not interpret the association between neglect and nonoral intake. As a result, the presence of neglect may be related to feeding limitations rather than swallowing deficits leading to nonoral intake.

More recently, increased systematic attention has been afforded sensory functions in swallowing and swallowing impairment.\(^5\)\(^-\)\(^10\) Continued emerging information and clinical observations suggest that impaired sensory functions may have a direct influence on swallowing functions. A better understanding of the role of sensory systems on swallowing function and impairment may lead to improved sensory-based interventions for dysphagia (see Clinical Corner 3-1).

**Issues of Unilateral versus Bilateral Hemispheric Lesions**

The issues previously raised regarding hemispheric contribution to swallowing control also raise the question of
whether such control is unilateral or bilateral. A traditional perspective is that patients with bilateral lesions often demonstrate the most severe and persistent dysphagia characteristics.\textsuperscript{11} Still, patients with unilateral hemisphere lesions may demonstrate dysphagia to varying degrees. Research using the technique of transcranial magnetic stimulation has suggested an interesting point of view on the hemispheric representation of swallowing function. Transcranial magnetic stimulation involves sending a magnetic current across the cranium over discrete hemisphere regions. These magnetic currents stimulate motor activity that is measured in various muscles by electromyography. This interesting work on the hemispheric control of swallowing function can be summarized as follows:

1. Swallowing motor functions are bilaterally represented in the hemispheres.
2. If the dominant hemisphere is impaired, a contralateral “backup” area may be available to facilitate recovery.
3. A form of cortical plasticity may occur over time, increasing the utility of the intact, nondominant hemisphere to control swallowing motor functions.
4. Bilateral strokes would result in the most tenacious dysphagias.\textsuperscript{12-15}

In some respects, this perspective is consistent with traditional clinical observations; bilateral strokes produce the most severe dysphagia, and many patients with unilateral strokes often recover the ability to swallow after a period of dysphagia.

### SWALLOWING DEFICITS IN HEMISPHERIC STROKE SYNDROMES

Several issues must be addressed when considering dysphagia secondary to hemispheric strokes. These issues may be simplified into two general considerations: location and extent of damage and functional consequences of the damage. These considerations are not mutually exclusive. Location and extent of the damage may be important in understanding sensory and motor impairments and in understanding the severity and potential for recovery based on unilateral versus bilateral lesions. In clinical practice, information on lesion characteristics often is not available at the time of the dysphagia evaluation. Therefore a strong reliance on the clinical examination of functional impairment after stroke may provide the best “road map” to understanding and perhaps predicting dysphagia characteristics. Table 3-1 provides a basic orientation to some of the functional impairments that may be clinically observed after impairment to various levels of the nervous system. At the hemisphere level, intent to swallow may be an important consideration. If the patient indicates such intent, a subsequent consideration would be motor initiation of the swallow. Patients with damage to premotor areas (e.g., supplemental motor cortex) may have generalized difficulty with motor initiation. The clinical picture may be that of a patient who holds a bolus in the mouth for an abnormally long period with associated movements that indicate the intent to swallow but without initiating a swallow (see Practice Note 3-1).

Patients with sensory deficits may demonstrate a variety of dysphagia characteristics, including retention of a portion of a bolus in the mouth, oropharynx, or hypopharynx with no attempt to clear the residue. These patients also may be more susceptible to aspiration of material into the upper airway as a result of the sensory deficit. Another category of sensory deficit may be seen in the patient with neglect. Such patients may not recognize material presented to one side of the swallowing tract. These patients may hold material in the mouth with no apparent intent to swallow, but in fact they are unaware of the material in the mouth.

Finally, patients with hemispheric stroke may have significant communication deficits or cognitive deficits that reduce their ability to relate to the clinical examiner the nature of the dysphagia complaints. Patients who are asleep, lethargic, have waxing and waning alertness, or difficulty
PRACTICE NOTE 3-1

The difference between intent to swallow and poor initiation of the swallow may be difficult to ascertain clinically. Some patients may hold food or liquid material in the mouth but not make any overt attempt to swallow. When this situation is encountered, one strategy is to observe differences in swallows when the patient self-feeds versus when the clinician provides the material to be swallowed.

Several years ago we encountered a young woman who had a severe stroke. Details of the stroke were sparse other than it was large and involved the frontal areas of the brain. She was nonverbal but did produce some vocalizations and demonstrated a very hypokinetic appearance. She was receiving all nutrition and hydration by gastrostomy tube. During the fluoroscopic swallowing study a clinician placed materials in the patient’s mouth by spoon. No overt reaction was noted to the placement of liquid or pudding material in the mouth. A colleague who was observing the examination suggested having the patient self-feed. We did not believe this was viable because of the paucity of spontaneous movement participating in the swallowing evaluation because of cognitive deficits present significant challenges to a valid evaluation of swallowing abilities (see Practice Note 3-2). Also, the inability to describe swallowing difficulties may delay or hinder clinical evaluation and implementation of rehabilitation strategies. Figure 3-2 depicts general hemispheric areas that may be associated with various sensorimotor functions associated with swallowing. The left hemisphere is shown for descriptive purposes only. Box 3-1 presents various swallowing characteristics that may be associated with sensorimotor deficits after hemispheric stroke.

A variety of swallowing deficits have been reported after hemispheric stroke. In general, hemispheric lesions (including both cortical and subcortical damage) contribute to many swallowing deficits (Box 3-2), including (1) poor initiation of saliva swallows (sometimes termed the dry swallow), (2) delay in initiation of the pharyngeal component of the swallow, (3) incoordination of the oral components of swallowing, (4) increased pharyngeal transit time and reduced pharyngeal constriction and clearing, (5) aspiration, (6) dysfunction of the pharyngoesophageal segment (cricopharyngeal muscle), and (7) poor relaxation of the lower esophageal sphincter. These collective observations indicate that hemispheric stroke can impair swallowing functions from the mouth to the stomach. Furthermore, a wide spectrum of swallowing deficits has been noted, ranging from impaired initiation of the swallow to poor transport of the bolus to aspiration into the airway. To date no report has emerged comparing specific sensorimotor stroke sequelae with specific swallowing impairments. However, the preceding list suggests that the array of potential swallowing deficits after demonstrated by this patient, but we placed a spoon in the patient’s hand and assisted her through the movements of filling the spoon with a liquid and then placing it in her mouth. The difference in swallow was dramatic. The woman swallowed the liquid material almost immediately. She demonstrated little residue and no airway compromise. This pattern was repeated for all materials presented in this manner. This simple change in feeding strategy was subsequently used in her daily rehabilitation program and she eventually returned to total oral feeding.

Refer to Video 3-1, A and B, on the companion Evolve site for an example of fluoroscopic swallowing differences in a single patient. This patient had a large right hemispheric middle cerebral artery stroke. In her initial fluoroscopic swallowing examination the clinician “fed” the patient barium contrast materials. A week later, the fluoroscopic examination was repeated because the reported results did not match the clinical profile. In this second examination we asked the patient to self-feed. The differences were dramatic.
stroke is extensive and may relate to the spectrum of post-stroke sensorimotor impairments (see Practice Note 3-3).

Dysphagia is highly prevalent in acute stroke, with estimates that well over 50% of all patients are affected. Early identification of dysphagia in acute stroke is a critical feature of clinical management as dysphagia is related to numerous health complications. Fortunately, the majority of acute stroke patients recover functional swallowing ability within the first 1 to 6 months after stroke, whereas swallowing problems develop in a small percentage of patients during the postacute period.\textsuperscript{16-18} These observations emphasize the importance of accurate identification and management of swallowing deficits in acute stroke patients. Furthermore, it is important to understand factors that might predict persistent swallowing problems beyond the acute recovery period. The importance of this perspective is highlighted by the observation that acute and chronic swallowing problems in stroke patients are associated with many complications, including increased length of hospitalization, dehydration, malnutrition, aspiration, chest infections and, in some cases, death.\textsuperscript{18-21} Furthermore, dysphagia during acute stroke is associated with poor long-term outcome, including death and an increased rate of institutionalization.\textsuperscript{22}

**Treatment Considerations**

Perhaps the most obvious statement about dysphagia in stroke is that it changes over time. From that perspective, dysphagia intervention strategies should also change over time. Table 3-2 presents clinical considerations and decisions that may affect treatment planning over time. Early in the course of a stroke, focus should be given to basic decisions such as the safety of oral feeding versus the need for nonoral feeding routes, the presence of comorbid conditions such as pneumonia (or other infections), malnutrition, dehydration, and the overall medical condition of the patient.

The acute stroke patient is at greatest risk for dysphagia and morbidities associated with dysphagia. The presence, or more accurately, the risk of dysphagia in acute stroke is best identified with screening programs (see previous discussion and Clinical Corner 3-2). In general, any patient who “fails” an early dysphagia screen should be thoroughly assessed during the early acute stroke period to confirm the presence and detail the characteristics of any dysphagia.
Recently I saw two patients within a short time frame who had similar histories and clinical presentations. Both patients were stroke survivors and at least 6 months past the stroke event. Both were deemed medically stable. Both depended on nonoral percutaneous endoscopic gastrostomy (PEG) feedings. And both had difficulty managing their oral secretions; they drooled and carried a towel to “mop up the problem.” Finally, neither patient had functional speaking ability, but both could vocalize and phonate simple vowels. These patients were referred for evaluation and treatment of pharyngeal dysphagia.

On the surface, this clinical presentation may make sense. However, the basic problem for both patients was not pharyngeal dysphagia, but rather oral apraxia. Admittedly, oral apraxia was quite severe in both patients. Both presented with a persistent open-mouth posture but in the absence of overt weakness within the facial or oral musculature. Both could close the mouth in response to intraoral sensory stimuli and both spontaneously swallowed, though infrequently. A key feature of the clinical examination in both cases was the absence of overt cranial nerve deficits. Also, each patient had the ability to close the mouth, but this was context dependent (e.g., they did not close the mouth on command, but when liquid was placed in the posterior mouth they did close and a spontaneous swallow was observed). As part of the clinical swallow examination, liquid was placed in the oropharyngeal area with a straw as a pipette. As this liquid trickled into the hypopharynx, we occasionally observed a swallow. Under endoscopic inspection we delivered additional liquid to the oropharynx in this fashion. I also learned that both patients protected their airway and that no residue remained after these volume-dependent swallows.

Based on these clinical and endoscopic findings I did not enroll these patients in therapy for pharyngeal dysphagia. The dysphagia was primarily the result of a severe oral apraxia that limited oral motor control for voluntary tasks, including swallowing.

I did make simple recommendations that I hoped would improve oral functions for feeding and oral control for swallowing. Because I observed intermittent spontaneous swallows when liquid was placed in the posterior part of the mouth, I recommended this technique in an attempt to stimulate improved oral swallow initiation. I suggested to the local therapist that if the frequency of spontaneous swallowing improved, she should vary the type and amount of material used in this fashion and gradually place the material more forward within the mouth. Later I heard from one of the local therapists that her patient had increased the frequency of spontaneous swallowing and was taking more oral intake. Sometimes success comes in small steps.

Refer to Video 3-2, A and B, on the Evolve site for endoscopic and fluoroscopic examples of a single patient who demonstrated significant oral apraxia in the presence of preserved pharyngeal swallowing function.

During the acute phase of stroke, patients are likely to demonstrate significant weakness contributing to reduced stamina and perhaps reduced mental status, including alertness and attention. These factors significantly limit any meaningful clinical (or other) evaluation of swallowing ability. Thus a conservative strategy is to observe the patient’s status and postpone any in-depth assessment or intervention until the patient is more alert and has better endurance. Acute stroke patients also are at risk for respiratory abnormalities. Respiratory abnormalities include basic weakness in expiratory muscles that might reduce cough effectiveness, increased episodes of oxygen desaturation, deviations in the respiratory rate, and alterations in the coordination between respiration and swallowing. Collectively, these respiratory deviations noted in acute stroke patients suggest an increased risk of aspiration of swallowed materials and pooled secretions and potential limitations in clearing aspirated secretions as a result of reduced cough efficiency. Given these potential risks, respiratory functions in the acute stroke patient should be evaluated as part of the comprehensive swallowing examination.

Pneumonia is noted in approximately 10% of acute stroke patients, with a higher prevalence if patients in the
intensive care unit are included.\textsuperscript{29} Pneumonia is a significant morbidity because it is related to both an increased number of hospital readmissions\textsuperscript{40} and short-term and long-term mortality.\textsuperscript{31} Causes of pneumonia in the poststroke patient are multifactorial; however, dysphagia, especially dysphagia accompanied by aspiration, is significantly related to the presence of pneumonia.\textsuperscript{19} In fact, dysphagia screening leading to early identification and treatment of swallowing deficits in acute stroke patients has been associated with a reduction in pneumonia rates.\textsuperscript{32-34}

The presence of dysphagia after stroke may contribute to pneumonia in various ways. Although the focus is often on aspiration of orally ingested food and liquid, aspiration of pooled pharyngeal secretions also may contribute to chest infection. Aspiration of secretions may be especially problematic in the acute stroke population because oral bacteria colonization is prominent in these patients.\textsuperscript{35} Patients dependent on tube feeding, specifically nasogastric tube feeding, may have a higher degree of bacterial colonization than patients who feed orally.\textsuperscript{36} In fact, at least one clinical research team has reported that stroke patients who were dependent on nonoral feeding (e.g., nothing by mouth) demonstrated higher rates of respiratory infections than did stroke survivors who were feeding orally.\textsuperscript{37-39} One implication of these findings is that reduced frequency of swallowing contributes to an increased risk of aspirating pharyngeal secretions in the presence of higher rates of bacterial colonization within the swallowing mechanism.\textsuperscript{40} This premise is supported by treatment studies demonstrating that swallowing therapy\textsuperscript{41} and strategies to improve oral hygiene\textsuperscript{42,43} reduce the incidence of pneumonia in stroke patients. Thus the dysphagia clinician should consider more than aspiration of food and liquid when providing swallowing interventions to patients after acute stroke.

Nutritional and hydration deficits are prevalent among stroke patients on admission and may worsen during hospitalization. On admission, the prevalence of nutritional deficits has been estimated at approximately 16%; this figure increases to 22% to 26% through discharge from acute care.\textsuperscript{44-46} Moreover, at least one study has identified an initial association between dysphagia and dehydration in acute stroke.\textsuperscript{40} In this study 53% of acute stroke patients demonstrated some degree of dehydration based on laboratory values (blood urea nitrogen/creatinine >15:1). Patients with dysphagia demonstrated greater dehydration compared to those without dysphagia. Furthermore, the degree of dehydration as measured by the laboratory value increased selectively among dysphagia patients during acute care. Though preliminary, these results place a strong focus on the hydration status of acute stroke patients, especially those identified with dysphagia.

Nutritional decline continues beyond acute care. The prevalence of nutritional deficits in stroke patients at admission to rehabilitation approximates 50%.\textsuperscript{47} At approximately 1 month after stroke, nutritional status begins to improve and continues to improve up to 4 months after stroke. In the acute stroke patient, nutritional deficits are not overtly linked to dysphagia.\textsuperscript{45,48} However, later during the rehabilitation period and thereafter, swallowing and feeding difficulties may contribute to the maintenance or increase in poor nutritional status.\textsuperscript{49} Still, some suggest that poor nutrition during the acute phase of stroke contributes to poor long-term functional outcomes.\textsuperscript{50} Nutritional evaluation and intervention are outside the scope of practice for most dysphagia clinicians. However, all dysphagia clinicians should be aware of the potential impact of swallowing and feeding abilities on nutritional status and participate in multidisciplinary health care teams that include nutritional specialists.

As the patient’s condition improves and more active rehabilitation is initiated (usually well within the first month after stroke), dysphagia treatment strategies also may change. One consideration is spontaneous resolution of dysphagia as the patient recovers from the effects of acute stroke. Although many stroke patients have some degree of

<table>
<thead>
<tr>
<th>TABLE 3-2 Treatment Considerations and Decisions for Dysphagia after Stroke*</th>
</tr>
</thead>
<tbody>
<tr>
<td>Considerations</td>
</tr>
<tr>
<td>-----------------</td>
</tr>
<tr>
<td><strong>Acute (0-1 Month)</strong></td>
</tr>
<tr>
<td>Most comorbid conditions</td>
</tr>
<tr>
<td>Resolving dysphagia</td>
</tr>
<tr>
<td>Nutrition/hydration</td>
</tr>
<tr>
<td><strong>Improving (1-6 Months)</strong></td>
</tr>
<tr>
<td>Patient more stable with better endurance</td>
</tr>
<tr>
<td>Comorbid conditions often under medical control</td>
</tr>
<tr>
<td>Feeding routes established for most</td>
</tr>
<tr>
<td>Malnutrition may still be a factor</td>
</tr>
<tr>
<td><strong>Chronic (After 6 Months)</strong></td>
</tr>
<tr>
<td>Feeding routes more established</td>
</tr>
<tr>
<td>Patients eating orally may have impaired swallow</td>
</tr>
<tr>
<td>Compensations that interfere with swallow</td>
</tr>
<tr>
<td>Impact of prior therapy</td>
</tr>
</tbody>
</table>

*Changing issues with time after onset.
recovery in swallowing ability, estimates of persisting dysphagia range from 11% to 50% at 6 months after stroke. During this period of improvement the patient with persistent dysphagia is likely to be engaged in active swallowing rehabilitation. By this time a decision about oral or nonoral feeding has already been established and comorbid conditions are often under medical control. Of importance to active dysphagia rehabilitation are various patient issues and the nature of the swallowing deficit. If the patient is able to participate in active rehabilitation and is motivated, direct and intense swallowing therapy is expected to produce significant benefit. Benefits from swallowing therapy extend beyond improved swallowing abilities to include reduced pneumonia rates and improved nutritional status.

Decisions about therapy techniques selection depend in large part on the specific dysphagia characteristics demonstrated by individual patients.

Chronic dysphagia is reported in some stroke survivors, although no study has documented the prevalence of dysphagia in stroke patients beyond 6 months after stroke. Typically, if the swallowing deficit persists beyond 6 months, it is considered chronic. Available reports indicate that stroke patients with chronic dysphagia can benefit from intensive therapy. Such therapies are typically active and directed at changing specific physiologic features of the swallowing deficit.

In summary, dysphagia is highly prevalent after stroke and may be related to pneumonia (and other infections), nutrition and hydration deficits, and other health complications. Dysphagia does resolve to varying degrees in the poststroke period, but the few estimates available suggest that up to 50% of stroke patients demonstrate some degree of persistent dysphagia. Dysphagia therapy has been shown to improve swallowing ability, reduce pneumonia rates, and improve nutritional status in stroke patients. Even stroke survivors with chronic dysphagia can experience functional benefit from intensive swallowing therapy.

**SWALLOWING DEFICITS IN DEMENTIA**

Another form of cortical impairment that can affect swallowing ability is the category of progressive diseases known as dementia. Several types of dementias have been described; the most frequent is Alzheimer’s disease. Other forms of dementia include dementia caused by cerebrovascular disease, Lewy body dementia, frontotemporal dementia, alcoholic dementia, and vitamin B12 deficiency, among others. Dementia typically involves progressive deterioration in cognitive abilities, including memory, judgment, abstract reasoning, and personality changes. Other cortical disturbances such as apraxia or aphasia might be noted.

Swallowing deficits are well documented in advanced dementia. A recent systematic review reported that across all forms of dementia the prevalence of swallowing difficulties ranged from 13% to 57%. Persistent weight loss may be the first indication that patients with dementia have a significant swallowing problem; however, weight loss may not be directly related to feeding or swallowing difficulties. As a result, such individuals are at significant risk for nutritional deficits that may further compromise their health status. Pneumonia is a common cause of death in patients with dementia. Although dysphagia, including aspiration, is associated with pneumonia in this population, it is not the only contributing factor and may not be the critical contributing factor.

General characteristics of swallowing deficits in dementia are listed in Box 3-3. Prominent on this list is the presence of oral-stage dysfunction. Certain oral aspects of swallowing are under volitional motor control. From this perspective, generalized cognitive impairments in dementia may contribute to deficits in volitional motor control and hence oral aspects of dysphagia. Oral aspects of dysphagia in patients with dementia may be characterized by lack of initiation of the swallow in which the patient holds food in the mouth, uncoordinated oral control of food and liquid, and/or delayed initiation of the oral component of the swallow. Each of these dysphagia characteristics contributes to prolonged mealtimes, which may put patients with dementia at nutritional risk from reduced food intake.

Although the majority of dysphagia information in dementia is derived from studies of patients in advanced stages of the disease, patients with mild-stage dementia also demonstrate feeding and swallowing deficits. One interesting approach to identification of dysphagia early in patients with Alzheimer’s disease was described by Sato et al. These investigators evaluated a variety of oral and feeding activities during daily life (e.g., lip and tongue movement, ability to rinse and gargle orally, storing food in the mouth, appetite, etc.) and reported that oral rinsing ability was the single factor most significantly associated with dysphagia. At one level this observation is logical as

**BOX 3-3 SWALLOWING DEFICITS SEEN IN PATIENTS WITH COGNITIVE DECLINE (DEMENTIA)**

- Unexplained weight loss*
- Oral-stage dysfunction*
- Pharyngeal-stage dysfunction
- Combined oral and pharyngeal dysfunction
- Minor aspiration
- Major aspiration
- Feeding limitations

*More commonly observed characteristics.
Successful oral care and swallowing for the patient with advanced Alzheimer’s disease is very much in the hands of the caregiver. I learned this firsthand by caring for my mother, who had Alzheimer’s disease. As speech-language pathologists, we are often in a position to offer insight into the process of these two activities by combining our knowledge of swallowing with our knowledge of dementia. The following anecdotal examples illustrate this point.

First, how may the sensory input of taste trigger an oral behavior? Good oral hygiene is critical, but what do you do when it is no longer feasible to use regular toothpaste, because it is often mismanaged and swallowed? Toddler toothpaste (designed to be safe if swallowed) is an option and a popular flavor is bubble gum. There were occasions when my mother accomplished the desired swish and spit after the predictable brushing action. However, months later, when giving her liquid Tylenol (also in bubble gum flavor) she remembered to successfully swish and spit.

Second, and this may be very case specific, we encountered the dilemma of trying to cue chewing and swallowing during a meal. Although the process was slow and laborious, it seemed sensible that this should be done slowly with verbal and tactile cues with each step and a pause between each bite to check for residual food in the oral cavity. However, I stunned the nursing assistant by trying to follow a successful swallow with another bite without pausing. For a time this was a successful strategy. Why did I try this? One of the four “A’s” of Alzheimer’s disease is apraxia. I recalled that it is at the point of transition that motor planning seems to break down. Therefore if we minimized the pause between successful swallows, an almost “automatic” second swallow of food followed.

AUTHOR’S NOTE: This practice note was provided by my former student and good friend, Dr. Nancy J. Haak, who cared for her mother with Alzheimer’s dementia in her home. In multiple conversations with Nancy, I learned much about the practical management of dysphagia in patients with dementia and felt it appropriate to share at least one example in this text. — MAC

BOX 3-4 EXAMPLES OF SWALLOWING AND FEEDING DEVIATIONS IN MILD-STAGE DEMENTIA

Swallowing Deviations
- Slow oral movement
- Slow or delayed pharyngeal response
- Overall slow swallowing duration

Feeding Deviations: Patients May Require the Following to Maintain Oral Intake
- Increased self-feeding cues (specifically related to food preparation or utensil use)
- Direct assistance with utensil use for food preparation or convenience
- Imitation of feeding behavior from the meal partner

Successful oral rinsing requires a degree of oral motor control, which may be impaired among patients with Alzheimer’s disease who have swallowing difficulties (see Practice Note 3-4). Box 3-4 summarizes salient findings regarding feeding and swallowing abilities in mild-stage dementia. These impairments are similar to, though not as severe as, those reported in more advanced stages of the disease. Specifically, patients with dementia demonstrate an overall slowing of the swallowing process from the oral aspects of food manipulation through the response of the pharynx accepting the bolus. This slowing of the swallowing process can have direct consequences for longer mealtimes and hence increase the risk of involuntary weight loss and associated declining nutritional status. In addition, slowing of the pharyngeal response in swallowing may reduce airway protection, resulting in an increase of coughing and choking behaviors during mealtimes.

In addition to overall slowness in the swallowing process, individuals with dementia frequently demonstrate self-feeding difficulties. Self-feeding difficulties may relate to numerous factors, including cognitive impairment, motor deficits such as weakness or apraxia, loss of appetite, and food avoidance. Consequences of self-feeding difficulties can include weight loss and associated nutritional decline as well as dependency for feeding. Dependency for feeding can contribute to dysphagia-related health problems, including pneumonia. 70-72 Self-feeding difficulties may be noticed in the mild stages of the disease and become more pronounced as the disease progresses. 73,74 For patients with self-feeding difficulties, clinicians or caregivers may need to offer increased verbal or environmental cues or provide direct assistance. Refer to Video 3-3 on the companion Evolve website for this text for an example of feeding difficulties in a single patient with primary progressive aphasia (PPA). PPA is a form of dementia in which language and communication abilities deteriorate initially followed by deterioration of other functions. This patient appeared to have a specific form of apraxia that influenced her use of eating utensils.

At the beginning of this lesson the premise was offered that different neurologic deficits contribute to different clinical presentations of swallowing deficits. At least one study has evaluated differences in feeding and swallowing abilities in patients with Alzheimer’s disease compared with patients with frontotemporal dementia. 75 As the name implies, frontotemporal dementia is often characterized by frontal lobe signs, including loss of insight, disinhibition, impulsivity, poor self-care, stereotypic behavior, and more. Conversely, Alzheimer’s disease is characterized by progressive memory deficits that may affect many tasks of daily life because of forgetfulness, disorientation, or
impaired executive functions. Ikeda et al. used caregiver questionnaires to evaluate eating behaviors in patients with frontotemporal dementia versus Alzheimer’s disease. They evaluated five categories: swallowing problems, appetite change, food preferences, eating habits (including table manners and stereotype behaviors), and other oral behaviors. In general, swallowing problems occurred less frequently than other limitations and the frequency of dysphagia did not differ among the types of dementia. However, swallowing problems tended to occur earlier in the course of disease progression in the patients with Alzheimer’s disease. Patients with frontotemporal dementia demonstrated more frequent changes in appetite. These patients were more likely to demonstrate increased appetite compared with reduced appetite in patients with Alzheimer’s disease. In addition, patients with frontotemporal dementia demonstrated more food preferences than patients in the Alzheimer’s group. Still, more than 20% of the patients with Alzheimer’s disease demonstrated increased preference for sweets and other taste-related changes. As might be expected from the basic profile, patients with frontotemporal dementia demonstrated more deviations in eating behaviors. Patients in the Alzheimer’s disease group did demonstrate longer meal durations, decline in table manners such as eating with hands, and a tendency to prefer eating at the same time each day. Finally, the category of “other oral behaviors” included observations such as overstuffing the mouth, eating nonedible objects, snatching any food item within reach, or vomiting, including self-induced vomiting. In general, patients with Alzheimer’s disease scored low on these behaviors with the exception of overfilling the mouth when eating. This is an interesting study in many ways. First, it details caregivers’ observations of eating and swallowing behaviors in different groups of patients with dementia. Second, it supports the basic premise that the characteristics of the underlying neurologic disease affect the clinical presentation of dysphagia. Finally, it provides at least an initial description of feeding and swallowing behaviors that may be used by dysphagia clinicians in evaluating swallowing and related behaviors in patients with dementia.

Treatment Considerations

Dementia is a progressive disease with no known cure. Dysphagia intervention for patients with any form of dementia should keep that focus and incorporate basic principles of quality of life, dignity, and comfort. Dysphagia treatment options for patients with dementia may range from simple environmental adjustments to the use of nonoral feeding sources. Depending on specific problems in individual patients, some potential treatment avenues may include special food preparations, diet restriction, enhanced taste and flavor, changing the mealtime environment, increased mealt ime supervision and cueing, or a variety of other behavioral or environmental changes to facilitate increased food and liquid intake. Direct behavioral therapy to change swallowing mechanics also may be indicated.

Feeding tubes are frequently recommended for patients with advanced dementia as a mechanism to maintain nutritional support and avoid dysphagia-related comorbid conditions. However, the available evidence on the benefit of feeding tubes for this population suggests that they do not reduce the risks of aspiration pneumonia, may not prevent further decline in nutritional status, may not prolong survival, and seem to have no impact on overall functional status. In fact, the American Geriatrics Society has issued a position statement that feeding tubes are not recommended for older adults with advanced dementia.

A recent survey of national databases (Minimum Data Set and Medicare Claims Files) indicated that most feeding tubes were placed in nursing home residents with dementia during acute hospitalization. The most common reasons for these hospitalizations included pneumonia, dehydration, and dysphagia. The 1-year mortality rate was 64%, with median survival of 56 days after tube placement. Patients with feeding tubes also had a significantly higher rate of health care use after tube insertion. These observations are nearly opposite the results of a survey completed by speech-language pathologists working in the area of dysphagia. In that survey, many respondents believed that percutaneous endoscopic gastrostomy (PEG) improved nutritional status and increased survival, and nearly 40% believed that PEG was the standard of care for patients with advanced dementia. However, the majority of respondents did not believe that tube feeding improved quality of life or functional status for these patients. A slightly more recent survey of speech-language pathologists indicated that misperceptions about tube feeding in advanced dementia were common but that clinicians with more experience demonstrated greater knowledge about tube feeding outcomes in this population. These misperceptions and lack of knowledge are not confined to speech-language pathologists. Pelletier evaluated dysphagia and feeding knowledge of certified nursing assistants working in nursing homes. Even though these professionals were actively participating in patient feeding activities, their knowledge of dysphagia and feeding was greatly limited. The results of this study and others suggest that focused education is vital in managing dysphagia and feeding limitations in patients with dementia. In fact, at least one study has demonstrated that educational programs for medical and allied health staff on end-of-life care and feeding management in patients with dementia resulted in a reduction in feeding tube placement in these patients. An additional study indicated that trained feeding assistance focusing on patients’ self-feeding ability, social stimulation during meals or snack periods,
and increased availability of choices for foods and liquids increased the daily intake of food and liquid in 90% of nursing home residents. Both feeding assistance and the availability of between-meal snacks resulted in increased oral intake. 86 Collectively, the available information suggests that (1) feeding tubes do not produce significant benefit to most patients with dementia, (2) they do not promote quality of life or compassionate care, (3) alternatives are available, and (4) education on the problems and intervention strategies can benefit t patients.

**SWALLOWING DEFICITS IN TRAUMATIC BRAIN INJURY**

Traumatic brain injury (TBI) typically results in diffuse neurologic deficits that affect several aspects of behavioral control. Various studies have indicated the prevalence of dysphagia in acute or subacute TBI ranges from 60% to more than 90%. 87-89 Oral-phase difficulties and pharyngeal-phase deficits are roughly evenly distributed within this population. 88 The primary factor related to the presence of dysphagia in these patients is the severity of neurotrauma assessed by clinical scales such as the Glasgow Coma Scale (GCS), the Rancho Los Amigos Scale (RLAS), or the Functional Independence Measure (FIM). 87-89 At least one study has suggested that the level of functional oral intake at admission to subacute rehabilitation as measured by the Functional Oral Intake Scale is one predictive component of return to total oral feeding in patients with TBI. 89 Another study reported that a level IV on the RLAS was required to initiate oral feeding and that a level VI on this scale was needed for return to total oral feeding. 87 Mandaville et al. 90 reported that increased age, low RLA score, presence of a tracheostomy tube, and aphonv combined to predict which patients would be discharged from acute care with a feeding tube. Recovery of swallowing function in TBI is good; most patients regain some degree of functional swallowing within the first 3 to 6 months after injury. 90,91,92 The severity of the initial injury emerges as a strong predictor of both the presence of swallowing deficits and time to recovery of functional swallowing ability.

Pneumonia is frequently seen in patients with TBI, especially early in the posttraumatic course of treatment. Hansen et al. 93 reported that 27% of patients admitted with a brain injury for early rehabilitation were being actively treated for pneumonia and that pneumonia developed in an additional 12% during rehabilitation. Clinical factors associated with the presence of pneumonia included severity of neurotrauma (GCS, RLAS), no oral intake on admission, and presence of tracheostomy tube or feeding tube. Woratyla et intubation versus in-hospital intubation as risk factors for pneumonia. Furthermore, Hui et al. 95 reported that for patients requiring mechanical ventilation, each additional day on the ventilator was associated with a 7% increase in the risk of pneumonia. Thus level of consciousness, tracheostomy tube, nonoral feeding, type of intubation, and number of days on mechanical ventilation all appear related to the development of early pneumonia in patients with TBI.

In addition to the effects of neurotrauma on swallowing ability in patients with TBI, swallowing may be affected by factors such as the need for tracheostomy and/or ventilator support, the presence of communicative and cognitive deficits, and the presence of physical deficits that may interfere with self-feeding ability. Tracheostomy tubes indicate some degree of compromise in the respiratory system, which is integral in the swallowing process. Also, these tubes may have a mechanical impact on swallowing physiology. However, at least one study has reported that the presence of tracheostomy tubes was not associated with increased rates of dysphagia or aspiration in trauma patients. 96 Patients with communicative or cognitive deficits present additional challenges to clinicians in the design of swallowing assessments or rehabilitation strategies because of patients‘ reduced understanding and interaction. Finally, physical deficits impose a degree of dependency for activities such as self-feeding. 97

**Treatment Considerations**

In as much as the deficits observed in TBI are multifactorial, the potential treatment strategies and techniques are also multifactorial. Cherney and Halper 98 provide a brief but excellent review of the roles of interdisciplinary team members that may be required in the management of dysphagia in patients with TBI. Standard intervention approaches included diet modifications, postural adjustments, feeding adaptations, and behavioral maneuvers and compensations. 99 In cases of severe injury with widespread comorbid conditions, alternate feeding routes may be indicated, especially in the early postinjury course. The good news is that many patients with dysphagia after TBI do regain the ability to eat by mouth with appropriate clinical intervention.

**SUBCORITICAL FUNCTIONS**

The basal ganglia are a group of cell bodies in the subcortical brain hemispheres that influence the quality of movement. Basal ganglia functions regulate tone (resting tension level of muscles) and steadiness of movement among other functions. Impairment to basal ganglia functions may create excessive tone and/or extra, unintended movements. Excessive tone may create delays in the initiation of
slowed movements, or a reduced amount of movement. Extra, unintended movements disrupt the smooth, coordinated nature of voluntary movement attempts. Movement disruptions may be seen as tremor, regular clonic movements, slow sustained postural interruptions (dystonias), or other unintentional movements superimposed on the normal resting state of muscle groups or during intended movements. Box 3-5 lists general swallowing problems that may be associated with various characteristics of basal ganglia deficits.

**BOX 3-5 GENERAL DYSPHAGIA CONSIDERATIONS IN PATIENTS WITH BASAL GANGLIA DEFICITS**

- Poor bolus control: involuntary movements
- Oral
- Oropharyngeal
- Residue from inefficient swallow
- Oral
- Oropharyngeal
- Pharyngeal
- Difference among swallow types
- Automatic versus intentional movements
- Severity dependent

Swallowing deficits in patients with PD are common and reflect the underlying motor impairments, the extent of the disease progression, and potentially the effects of medications. Miller et al. reported dysphagia in 84% of a sample of 137 adults with PD; 23% demonstrated severe dysphagia and could not complete a 150-mL water swallowing task. These prevalence data may be lower than the true clinical picture because patients, especially those in the earlier, milder stages of the disease, do not reliably report swallowing difficulties. For example, Kalf et al. reported an objective (clinician assessed) prevalence of dysphagia of 82% versus a subjective (patient report) prevalence of 35% in community-dwelling PD patients. In general, oropharyngeal swallowing deficits may result from poor bolus control caused by involuntary movements or from residue or misdirection of the bolus from an inefficient, possibly weakened swallow (see also Clinical Corner 3-3). In addition, an overall slowness characterizes swallowing deficits in patients with PD that may reflect the degree of underlying bradykinesia. In addition to the motor component of PD, sensory deficits may contribute to dysphagia and related difficulties. Hammer, Murphy, and Abrams reported abnormal airway somatosensory functions and increased oropharyngeal residue in PD patients compared with healthy controls. Moreover, these investigators described a positive correlation between sensory thresholds and swallowing impairment. These sensory deficits may be based in peripheral sensory nerve changes in the pharynx associated with the disease and may contribute to aspiration of saliva and perhaps other liquids in this population. Furthermore, these sensory limitations may contribute to underreporting of dysphagia symptoms by patients with PD. Box 3-6 lists some of the oropharyngeal swallowing-related deficits in patients with PD.

Drooling, in some contexts termed sialorrhea, is a common problem for patients with PD and may be related to the presence and severity of dysphagia. Sialorrhea in PD may result from a combination of sensory impairment and reduced frequency of spontaneous swallowing resulting in salivary retention. Results from preliminary studies have suggested that patients with diurnal sialorrhea are at increased risk for silent aspiration, which may, in turn, increase their risk for respiratory infections and subsequent death. These risks are higher in later stages of the disease and in patients with severe sialorrhea.

Swallowing deficits in PD extend beyond the oral and pharyngeal components of the swallowing mechanism. Gross et al. describe impaired coordination between swallowing and respiration that may contribute to reduced airway protection during swallowing. Moreover, various esophageal abnormalities have been reported, including delayed transport through the esophagus, esophageal stasis, abnormal contractions, and lower esophageal

**SUBCORTICAL FUNCTIONS AND SWALLOWING IMPAIRMENT: PARKINSON’S DISEASE**

Parkinson’s disease (PD) is a slowly progressive disease of the basal ganglia. The key problem is impairment in the execution of voluntary movement. The classic features of PD include resting tremor, bradykinesia, and rigidity. The cause of this disease is essentially unknown, but the immediate cause for the motor changes is the depletion of the neurochemical dopamine, which results in impaired basal ganglia functioning during voluntary movements. These changes may also result from long-term use of certain medications or may be part of more encompassing degenerative diseases that can influence basal ganglia performance.

Patients with PD may present with a variety of interrelated clinical signs. They may demonstrate slowness in cognitive tasks and in some cases a form of dementia. As the disease progresses, they may show a masklike face that appears expressionless. They often demonstrate a characteristic dystarthis, impaired writing (micrographia), changes in body posture and gait, and other potential changes associated with reduced movement ability or instability. The progression of PD varies among patients, and no cure currently exists for PD. Medical management consists primarily of medications, although recent efforts have described surgical approaches to management.
Years ago I saw a patient with PD for whom a feeding tube was being considered. I do not recall the stage of the disease, but he was nonambulatory outside his home, had some obvious degree of rigidity, and presented with a significant dysarthria (likely a 4 or 5 on the Hoehn and Yahr functional rating scale). I do remember that his wife was pleading with me and the radiologist to recommend that this patient could continue oral feeding, even if a feeding tube had to be placed for nutritional support. He had already had one episode of pneumonia, which prompted his referral for swallowing evaluation. Furthermore, the wife insisted that her husband could drink milkshakes at home with no difficulty.

Initially, the fluoroscopic swallow evaluation incorporated small volumes (5 mL or less) of thin liquid, thickened liquid, and pudding material provided to the patient by spoon. As expected, we noted poor oral control with material entering the pharynx before the airway was closed. We also noticed residue that increased in amount as the thickness of swallowed material increased. Given the wife’s report of successful milkshake drinking at home, we provided the patient with a cup of nectar-thickened liquid and a straw. To our surprise, his swallow improved dramatically under this condition. We observed no aspiration and only a small amount of residue once the sequence of multiple swallows was completed. This patient continued to take oral nutrition supplements by mouth for total nutrition for a short period. Even after a feeding tube was placed, he was able to continue drinking milkshakes by straw.

Critical Thinking
1. How would you explain the difference in swallowing performance based on straw drinking versus small volumes taken by spoon provided by the examiner? Consider neurologic, swallow mechanics, and context variables in your discussion.
2. Do you think this distinction may be specific to patients with PD or might other patients respond in a similar manner?
3. When do you think it is appropriate to evaluate swallowing abilities in patients diagnosed with PD? Refer to Video 3-4 on the Evolve website for an example of how different swallowing strategies and different bolus volumes may affect swallow function in a patient with PD. The initial swallow is a thick liquid presented by the clinician from a spoon. Subsequent swallows are taken sequentially by the patient with a straw.

<table>
<thead>
<tr>
<th>BOX 3-6 OROPHARYNGEAL SWALLOWING DEFICITS IN PATIENTS WITH BASAL GANGLIA DEFICITS (PARKINSON’S DISEASE)</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Oral Stage</strong></td>
</tr>
<tr>
<td>• Lingual tremor</td>
</tr>
<tr>
<td>• Repetitive tongue pumping*</td>
</tr>
<tr>
<td>• Prolonged ramplike posture</td>
</tr>
<tr>
<td>• Piecemeal deglutition</td>
</tr>
<tr>
<td>• Velar tremor</td>
</tr>
<tr>
<td>• Buccal retention*</td>
</tr>
<tr>
<td><strong>Pharyngeal Stage</strong></td>
</tr>
<tr>
<td>• Vallecular retention*</td>
</tr>
<tr>
<td>• Piriform sinus retention</td>
</tr>
<tr>
<td>• Impaired laryngeal elevation*</td>
</tr>
<tr>
<td>• Airway (supraglottic) penetration</td>
</tr>
<tr>
<td>• Aspiration</td>
</tr>
<tr>
<td><strong>Sensory Deficits (Elevated Threshold for Sensation)</strong></td>
</tr>
<tr>
<td>• Pharyngoesophageal segment dysfunction</td>
</tr>
</tbody>
</table>

*More commonly observed characteristics

abnormalities. Patients with PD have been reported to demonstrate problems farther along the digestive tract—gastroparesis and various defecatory dysfunctions. Again, these irregularities may be related to the movement disorder or to the influence of some of the medications used to treat the disease. Still, dysphagia clinicians should at least discuss the entire spectrum of gastrointestinal functions in evaluating dysphagia in patients with PD.

It is important to remember that patients with PD must cope with a widespread assortment of daily problems resulting from the disease and, at times, from the treatments for the disease. These deficits extend beyond the swallowing mechanism and may affect related acts such as food shopping, preparation of meals, and self-feeding activities. Thus dysphagia in patients with PD and associated daily activities may contribute to increased patient and caregiver burden. In the absence of appropriate support systems, these dysphagia-related impairments could have a direct, and potentially negative, influence on the nutritional and health status of individual patients.

**Treatment Considerations**

Clinical research on the effectiveness of dysphagia therapy for patients with PD is limited. In fact, a systematic reviews by Baijens and Speyer and van Hooren et al. identified only 16 and 12 articles respectively describing rehabilitative, surgical, pharmacologic, or other therapies for swallow difficulties in PD. Not surprisingly, most articles reported some degree of positive benefit from their particular interventions. In fact, it is conceivable that a variety of interventions may improve some aspects of swallow
function in patients with PD. For example, Felix et al. reported improved swallowing of water and to a lesser extent biscuits after a 2-week period of performing the effortful swallow technique with adjunctive biofeedback. Athukorala et al. also used adjunctive biofeedback but employed a novel application in which patients focused on a skill that required spatiotemporal coordination rather than just increased strength of swallowing. Timing aspects of swallowing improved immediately following therapy and were maintained for 2 weeks following therapy. El Sharkawi et al. reported that some swallowing variables improved after 1 month of Lee Silverman Voice Treatment (LSVT). LSVT is a well-known therapy for speech and voice improvement in patients with PD. This study examined the cross-system effect of LSVT on swallowing performance. Finally, Pitts et al. reported that 4 weeks of expiratory muscle strength training improved both voluntary cough and some swallowing parameters. Collectively, these studies represent a wide range of behavioral interventions both in terms of the focus and outcomes of therapy. However, each intervention may be appropriate and helpful to select individual patients with dysphagia attributable to PD. In addition, dysphagia clinicians are advised to remember that medical and surgical interventions may be appropriate for certain patients. From an evidence-based perspective, the available literature reflects small numbers of patients with generally weaker study designs. As the clinical sciences mature, the expectation is that clinicians and patients will benefit from more rigorous knowledge on the effectiveness of various dysphagia interventions for patients with PD.

As with all dysphagias, treatment planning interacts with an understanding of the underlying mechanisms contributing to the dysphagia. In addition, because PD is a progressive disease, intervention strategies are expected to change over time. Finally, some evidence suggests that medications may have a positive effect on swallowing function in patients with PD, however, this benefit may not extend to all patients or to some aspects of swallow function. Because medications tend to work in time cycles, it may be important to time meals in relation to the maximum beneficial effect of medications. Finally, Table 3-3 summarizes intervention strategies recommended by Yorkston et al. that may be appropriate for patients with PD. Although clinicians should not limit treatment options to those listed in the table, the recommendations do reflect the changing nature of dysphagia in PD over time and represent a range of potential interventions from patient counseling and education to modifying swallowing activity to adjusting diets. As such, this information may serve as a general guide to dysphagia clinicians with common sense suggestions at various severity levels of PD.

<table>
<thead>
<tr>
<th>Presenting features</th>
<th>Normal Swallow</th>
<th>Early Swallowing Problems</th>
<th>Moderate Swallowing Disability</th>
<th>Severe Swallowing Disability</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>No observable changes</td>
<td>Reduction in pharyngeal peristalsis</td>
<td>Pharyngeal peristalsis worsens</td>
<td>Aspiration both during and after swallow</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Repetitive rocking motion of the tongue</td>
<td>Delay in swallowing reflex</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>Cricopharyngeal dysfunction</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>Laryngeal closure during swallowing may be inadequate</td>
<td></td>
</tr>
<tr>
<td>Intervention</td>
<td>Monitor weight</td>
<td>Provide counseling to bring swallowing under voluntary control</td>
<td>Introduce aids and devices to promote independence</td>
<td>Teach chin-tuck swallowing</td>
</tr>
<tr>
<td>Answer questions</td>
<td>Monitor weight</td>
<td>Increase sensory input</td>
<td>Switch to soft diet</td>
<td></td>
</tr>
<tr>
<td>Coordinate eating with drug cycle</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Recommend small, frequent, highly nutritious meals</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

(From Yorkston KM, Miller RM, Strand EA: Management of speech and swallowing in degenerative diseases, Tucson, AZ, 1995, Communication Skills Builders.)
BRAINSTEM FUNCTIONS

The brainstem is much like a junction box. Here the major ascending sensory tracts receive input from the head and neck region by way of the cranial nerves. The head and neck musculature also receives motor innervation from the upper motor neurons of the corticobulbar system. These upper motor neurons synapse with the motor components of the individual cranial nerves, which function as lower motor neurons. Thus damage to the brainstem typically results in sensory deficits to the head and neck region in addition to motor deficits associated with both upper and lower motor neuron damage. The first of these is characterized by spastic weakness and associated movement impairments, whereas the second is characterized by flaccid weakness and associated movement impairments. The term alternating hemiplegia is often applied to this pattern of motor impairment to describe flaccid weakness on one side of the body (head) and spastic weakness on the contralateral side (body). In simple terms, the “level” of brainstem deficit is noted by the cranial nerve level of flaccid weakness. Thus, facial alternating hemiplegia indicates a flaccid weakness in the facial or seventh cranial nerve with spastic weakness on the contralateral side (body). In more technical terms, it does offer an overt description of the impairment in swallow physiology observed in these patients. Box 3-7 summarizes features of the incomplete swallow often seen in patients with dysphagia subsequent to brainstem stroke.

A careful assessment of the components of dysphagia and related deficits is mandatory in this group of patients. For example, the patient requiring tracheostomy for respiratory support presents a different clinical profile than does the patient who does not require tracheostomy. The patient with minimal cranial nerve deficits may have better physiologic support for rehabilitative efforts than the patient with multiple cranial nerve deficits. And the nonambulatory patient presents different challenges than the patient who can walk assisted or unassisted.

In the acute poststroke phase, intervention tends to be more cautious with a prophylactic component. At this point the patient may be at greatest risk for pulmonary complications from inappropriate oral intake. Depending on the severity of neurologic impairment and the overall health status of the patient, treatment strategies at this

<table>
<thead>
<tr>
<th>BOX 3-7 PHARYNGEAL SWALLOWING DEFICITS IN PATIENTS AFTER BRAINSTEM STROKE</th>
</tr>
</thead>
<tbody>
<tr>
<td>• Absent or delayed pharyngeal response</td>
</tr>
<tr>
<td>• Reduced hyolaryngeal excursion</td>
</tr>
<tr>
<td>• Reduced oropharyngeal constriction</td>
</tr>
<tr>
<td>• Reduced pharyngeal constriction</td>
</tr>
<tr>
<td>• Reduced laryngeal closure</td>
</tr>
<tr>
<td>• Reduced pharyngoesophageal segment opening</td>
</tr>
<tr>
<td>• Brief swallow event</td>
</tr>
<tr>
<td>• Generalized incoordination (including respiration)</td>
</tr>
</tbody>
</table>

Brainstem Functions and Swallowing Impairment

Swallowing deficits subsequent to brainstem stroke provide a good example of the relation between neurologic deficits and dysphagia. In general, dysphagia in brainstem stroke involves two aspects: incoordination presumably related to disruption of the “swallowing center” and weakness resulting from damage to the corticobulbar system (sensory deficits also may be present). The collective effects of these deficits often are manifest clinically as incoordination among “stages” of swallowing and between swallowing and respiration, as well as weakness in one or more of the muscle groups innervated by the corticobulbar system (velum, pharynx, larynx, pharyngoesophageal segment). The resulting swallow has been described as the incomplete swallow. Although incomplete swallow is not a specific term, it does offer an overt description of the impairment subsequent to brainstem stroke. In general, dysphagia in brainstem stroke recover some degree of swallow function over time. Likewise, the clinical presentation of dysphagia and comorbid conditions varies considerably. Given these perspectives, treatment approaches to dysphagia in the patient who has survived a brainstem stroke are symptomatic and change over time.
stage may range from nothing (monitoring recovery) to passive sensorimotor activities (oral hygiene and movement exercises) to more active swallowing efforts involving compensatory maneuvers (postural adjustments, changes in the swallow behavior, etc.) or even intensive rehabilitation exercises.

Because recovery facilitates an overall improvement in the patient’s health status, dysphagia intervention may be more direct and aggressive. At some point the need for continuation of tracheostomy tubes should be addressed. Direct and intensive swallowing rehabilitation has been effective in facilitating return to oral feeding in chronic patients. 55-57 Although limited, clinical research has suggested that therapy approaches focused on increasing strength and coordination of swallowing are likely to improve swallow function. The key for the dysphagia specialist is interaction with medical and other rehabilitative specialists to understand the patient’s larger health status picture and selection of treatment strategies consistent with the patient’s global needs and still provide the potential for improved swallowing function.

The Role of the Cerebellum in Swallowing

The cerebellum is adjacent to the brainstem and is located posterior and slightly superior to most brainstem structures. The role of the cerebellum in the control of swallowing is poorly understood. This structure does appear to play a role in swallow activity; several functional imaging studies have demonstrated activation, often bilateral activation, in the cerebellum on volitional swallowing. 136-138 A recent review of literature from 1980 forward concluded that the cerebellum likely has some role in modulating swallowing and can contribute to dysphagia when damaged. 139 From a clinical perspective, cerebellar damage results in unsteadiness (ataxia), intention tremor (tremor that is exaggerated at the initiation of movement), and hypotonia (low muscular tone). When present in the swallowing mechanism, these movement deficits are expected to impair coordinated swallowing functions. Motor unsteadiness and weakness resulting from cerebellar damage may contribute to difficulty in controlling a bolus, directing that bolus in a timely fashion, and residue from reduced swallowing effort. However, given the location of the cerebellum, clinicians must be vigilant of brainstem (cranial neuropathy and central pattern generator) contributions to any dysphagia resulting from primary damage to the cerebellum (see Practice Note 3-5). Video 3-7 on the companion Evolve website presents an endoscopic swallowing examination of a patient with cerebellar deficit who demonstrates tremor that contributes to poor oral control of a liquid bolus with subsequent aspiration.

PRACTICE NOTE 3-5
Our dysphagia treatment team received a referral to assess a young woman who had been on a feeding tube for 5 years. Her history indicated that a viral infection had interfered with her blood clotting ability, which resulted in a large midcerebellar stroke. Though the stroke did not directly affect the brainstem and she demonstrated no cranial nerve deficits, she did have a severe and persistent dysphagia and respiratory difficulties. She required months of hospitalization and ventilation support for breathing. Subsequently, she completed years of physical and swallowing rehabilitation. When we saw her, we dubbed her “the girl with no swallow.” A brief fluoroscopic video of her swallow pattern can be found on the Evolve website (Video 3-5). Her pattern was to drop the base of her tongue to allow material to “fall” into the hypopharynx. Once at the level of the piriform recesses (entrance to the esophagus) she immediately “reversed gears” and regurgitated the material into her mouth. This pattern was repeated until she was forced to breathe and the material was expectorated. In essence, she had lost any form of a functional swallow. The good news is that after intensive swallowing rehabilitation she was able to regain her swallow function and returned to a relatively normal oral diet.

LOWER MOTOR NEURON AND MUSCLE DISEASE

Lower motor neurons proceed through the body and connect with muscles at the myoneural junction. Deficits to the peripheral nerves or the myoneural junction produce flaccid weakness. However, myoneural junction deficits demonstrate significant deterioration of motor function with use but recovery with extended rest.

The end points in the sensorimotor chain of events are the muscle and sensory end organs. Motor impairments at the muscle level are termed myopathies. These are characterized by a severe flaccid weakness within the affected muscle groups. Sensory loss may come in many forms, resulting from both neurologic and nonneurologic processes. Reduction or loss of tactile sensation is considered particularly important in swallowing problems because it may lead to unawareness of residual food along the swallowing mechanism or it may contribute directly to aspiration of food and liquid materials into the airway.

Lower Motor Neuron Functions and Swallowing Impairment

Amyotrophic lateral sclerosis (ALS) is one disease that reflects the relation between lower motor neuron impairment and dysphagia. ALS, sometimes referred to as Lou
**Gehrig’s disease or motor neuron disease,** is a progressive degenerative disease of unknown cause. The clinical presentation is progressive weakness; approximately 30% of patients show the initial effects of this disease in the corticobulbar musculature. \(^{140}\) When present, corticobulbar deficits contribute to a significant and progressive dysphagia.

Neurologic deficits in ALS are not confined to the lower motor neurons of the peripheral nervous system. Central nervous system structures also are involved. As a result, the motor deficits in ALS are mixed—involving both flaccid (lower motor neuron) and spastic (upper motor neuron) weakness. The mixture of flaccid and spastic weakness may be seen in the musculature of the swallowing mechanism, in the respiratory musculature, and throughout the remainder of the body. ALS is progressive and terminal, and although many patients survive for longer than 5 years, the majority do not. \(^{140,141}\) Substantial variability in progression rates exists among individuals. Respiratory failure is a common cause of death. Available research suggests that the different subtypes of ALS do not progress differentially. \(^{142}\)

In addition to dysphagia, individuals with ALS experience movement difficulties with the arms and legs, dysarthria, respiratory decline from chest muscle weakness and, in some cases (though rare), cognitive changes (including emotional lability and dementia). Obviously the impact of this disease on all aspects of daily functions is severe. These factors certainly are considered in planning any rehabilitative efforts, including swallowing rehabilitation.

Swallowing deficits are progressive and widespread. As might be expected, they reflect a weakness across the muscle groups used to prepare and transport a bolus. Early in the course of the disease, dysphagia may be characterized by oral limitations resulting from lingual weakness. \(^{143,144}\) In fact, lingual weakness has been associated with survival time in ALS in at least one study \(^{145}\) and combined with respiratory measures may be a good indicator of a patient’s ability to take oral food and liquid. \(^{146}\) In addition to poor oral transport of a bolus, patients with ALS, even those with no bulbar symptoms, demonstrate pharyngeal residue. \(^{147}\) This observation may be related to early, undetected weakness in pharyngeal muscles. Solazzo et al. \(^{148}\) identified reported manometric irregularities in the pharynx and upper esophageal sphincter in the absence of fluoroscopic abnormalities in 10 patients with ALS. These findings suggest that weakness is present in swallowing musculature prior to clinical or fluoroscopic recognition of dysphagia. However, pharyngoesophageal segment opening and laryngeal excursions may demonstrate relative maintenance even in advanced dysphagia. \(^{147}\) As might be expected, respiratory aspects of swallowing are negatively affected in ALS. Nozaki et al. \(^{149}\) reported that swallow apnea, or hypopnea, was increased in patients with ALS and that patients with severe respiratory limitations or presence of aspiration on fluoroscopic swallow examination presented the longest apnea durations. General considerations for dysphagia are listed in Box 3-8, and specific dysphagia characteristics are presented in Box 3-9. In general, these deficits reflect limitations in oral bolus control, reduced ability to transport the bolus with resulting residue, and reduced airway protection. Because lingual weakness is an early aspect of dysphagia in ALS, it is not surprising that speech production also is affected. In fact, speech and swallow functions in ALS tend to show a highly related course of deterioration.

Early in the disease course, no significant dysphagia may be reported. As weakness in the swallowing mechanism progresses, patients may have difficulty chewing solid food, loss of food or liquid from the lips, and food-specific difficulties. This may cause patients to begin to reject specific foods or to alter their diet or chewing or swallowing

---

**BOX 3-8  GENERAL DYSPHAGIA CONSIDERATIONS IN PATIENTS WITH ALS AND ASSOCIATED SENSORIMOTOR DEFICITS**

- Oral control of bolus
- Perioral weakness
- Lingual weakness
- Reduced transport
- Velar leak
- Reduced tongue pump
- Reduced pharyngeal contraction
- Residue
- Airway protection
- Bradykinesia
- Residue
- Respiratory limitations
- Increased swallow apnea

ALSO, Amyotrophic lateral sclerosis.

**BOX 3-9  OROPHARYNGEAL SWALLOWING DEFICITS SEEN IN PATIENTS WITH ALS**

**Oral Stage**
- Leakage
- Mastication
- Bolus formation
- Bolus transport
- Residual pooling

**Pharyngeal Stage**
- Nasophasyngeal regurgitation
- Valleculae pooling
- Piriorm sinus pooling
- Airway spillage
- Ineffective airway clearance
- Shortness of breath

ALSO, Amyotrophic lateral sclerosis.
mechanics (see Practice Note 3-6). As the disease progresses further, patients need more extensive diet modifications and risk rapid weight loss, leading to nutritional decline. This situation, perhaps combined with the loss of a positive social environment surrounding mealtimes, may lead to the decision to use an alternate feeding source. Initially, patients may be able to continue some oral feeding, but at some point total reliance on alternate feeding sources may occur. Table 3-4 summarizes a variety of intervention strategies suggested by Yorkston et al. 129 across various stages of severity in ALS. Jenkins and colleagues 150 summarize the evidence for symptomatic treatments in ALS.

PRACTICE NOTE 3-6

A woman in her late 50s was referred for speech and swallow evaluation by her neurologist. Roughly 18 months before the evaluation she began to have speech difficulties. These were progressive, and roughly 5 months before the evaluation she noticed increased difficulty swallowing.

At the time of the clinical evaluation she was able to take all foods orally but she was avoiding “heavier” foods such as certain meats. She also engaged in swallow compensations, including cutting any masticated food into small pieces and using liquids to “wash” heavier food down when she ate them. She also reported difficulty controlling oral secretions, with resultant drooling day and night.

On clinical examination this woman demonstrated a mixed dysarthria. The tongue presented with bilateral fasciculations but other cranial nerves were grossly intact. Her score on the Mann Assessment of Swallowing Ability was 166 of 200, indicating moderate dysphagia. Her score on the speech scale of the ALS Severity Scale was 6, indicating the need to repeat some messages, and her score on the swallowing subscale was 7, reflecting diet changes.

Endoscopic and fluoroscopic swallowing examinations are presented in Video 3-8, A and B on the Evolve website. On endoscopic examination this patient demonstrates basic simple and simple movement but impairment on rapid and sequential movements. Still, her swallow abilities seemed functional. On fluoroscopic examination, she demonstrated a pattern of slowness with possible weakness, but she again gave the impression of a functional swallow.

I saw her again 2 months later. At this point she demonstrated little clinical change, although her MASA score had lowered to 154 with noted changes in tongue function and increased coughing during meals. The patient was having obvious difficulty coping with the apparent diagnosis and indicated a desire for no further clinical follow-up. These wishes were respected and she has not returned for additional evaluation or clinical assistance or advice.

They conclude that few treatments alter the course of the disease, but that many symptomatic treatments can have a positive influence on a patient’s quality of life. Noninvasive ventilation has been shown to support respiratory function, improve quality of life, and extend survival by approximately 7 months. Katzberg and Benatar 151 concluded that in the absence of strong evidence, the best available evidence suggested a survival advantage with improved nutrition of gastrostomy tube feedings for some patients with ALS. However, they caution that their findings are tentative. Furthermore, a pair of patient-oriented studies by Stavroulakis et al. 152,153 indicated that many patients preferred to delay feeding tube placement perhaps until swallowing difficulties reached a critical point, but that patient education both in the hospital and the community helped with the transition from oral to tube feeding. Finally, the role of exercise is still not clear for patients with ALS. Proponents of exercise suggest that especially in the early stages of the disease exercise benefits patients both physically and psychologically. Advocates indicate that exercise may slow disease deterioration resulting from disuse. 154 However, a Cochrane review by Dal Bello-Haas and Florence concluded that no evidence existed to claim benefit from exercise for patients with ALS. Likewise, no evidence existed that exercise was harmful to these patients. Based on the evidence (or lack thereof), the best clinical advice might be to evaluate the needs and motivation of each patient when considering an exercise approach to dysphagia in ALS.

Muscle Diseases and Swallowing Impairment

A variety of pathologic conditions may have a negative influence on muscles related to swallowing function. These diseases typically result in weakness in muscle groups that contribute to dysphagia. Examples of disease processes that might impair peripheral muscle function (in some cases including the peripheral nerve) include polyneuropathy, myasthenia gravis (MG), polymyositis, scleroderma, systemic lupus erythematosus, and dystrophy. Unless working as part of a specialized health care team, the typical dysphagia specialist does not encounter large numbers of patients with these disorders or diseases. However, it is important to recognize the potential impact of each condition on swallowing function and to be able to differentiate other causes of dysphagia from these clinical conditions. From that perspective, each of these muscle diseases with the potential to affect swallowing function is discussed briefly in relation to dysphagia characteristics.

Polyneuropathy

Literally meaning “pathology to many nerves,” polyneuropathies may result from many sources. Systemic diseases such as diabetes can result in polyneuropathies, as can other...
pro cesses that affect peripheral nerves. Perhaps most common to dysphagia, and often forgotten, is the peripheral nerve damage that can result from radiotherapy in the treatment of head and neck cancer. These patients have fibrosis in tissue as well as nerve deficits in the affected areas. Weakness in peripheral nerves innervating the swallowing musculature contributes directly to weakness in the muscles used for chewing and swallowing. Polyneuropathies also may result in sensory deficits with resulting effect on the ability to safely ingest food and liquid. Guillain-Barré syndrome is one example of a neurogenic polyneuropathy in adults. Nearly all patients with Guillain-Barré syndrome have some degree of dysphagia. Orlikowski et al.\textsuperscript{157} reported the reduced tongue strength was associated with dysphagia and respiratory limitations in patients with Guillain-Barré. Most patients recovered swallowing functions to varying degrees, but those with more severe dysphagia later in the disease tended to have persistent complaints.

**Myasthenia Gravis**

MG is a disease process in which the neurotransmitter substance between motor nerves and muscles is depleted with use. In this regard, initial movements (such as chewing) are often intact or at least at their strongest at the beginning of movement (such as a meal). With repeated use the muscles fatigue into a flaccid weakness. Thus any swallowing activity that requires sustained or repeated movement (i.e., most of them) results in fatigue and reduced function. Colton-Hudson et al.\textsuperscript{158} described dysphagia characteristics in 20 adults with MG. These investigators reported oral and pharyngeal deficits in all patient, and approximately 30\% demonstrated aspiration. In addition, Linke et al.\textsuperscript{159} reported that esophageal transit often is compromised in MG. Thus patients with MG may present with dysphagia characteristics reflecting weakness along the entire course of the upper swallowing mechanism. Finally, Warnecke et al.\textsuperscript{160} used a fiberoptic endoscopic examination of swallowing (FEES) to evaluate the immediate effect of the Tensilon test. Injection of Tensilon into a symptomatic patient with MG reduced symptoms within a short time. These authors reported that the combination of the FEES examination and the Tensilon test represents a clinical tool useful in the early diagnosis of MG-related dysphagia. One additional factor merits consideration. Available literature contains multiple case reports in which dysphagia was the initial presenting symptom of what eventually was diagnosed as MG.

---

**TABLE 3-4 Summary of Swallowing Interventions in ALS**

<table>
<thead>
<tr>
<th>Presenting features</th>
<th>Early Swallowing Problems</th>
<th>Dietary Consistency Changes</th>
<th>Unable to Meet Needs Orally</th>
<th>Salivary Problems</th>
</tr>
</thead>
<tbody>
<tr>
<td>Presenting features</td>
<td>Solid foods difficult to eat</td>
<td>Weight loss</td>
<td>Decline in calorie intake</td>
<td>Complaints of too much saliva</td>
</tr>
<tr>
<td></td>
<td>Longer mealtimes</td>
<td>Chronic dehydration</td>
<td>Decline in fluid intake</td>
<td>Complaints of drooling</td>
</tr>
<tr>
<td></td>
<td>Need for smaller bites</td>
<td>Loss of enjoyment</td>
<td>Food spillage from mouth</td>
<td></td>
</tr>
<tr>
<td><strong>Intervention</strong></td>
<td>Use chin-tuck position</td>
<td>Change to soft diet</td>
<td>Insert PEG or insert nasogastric tube or insert intermittent orogastric tube</td>
<td>Maintain adequate hydration</td>
</tr>
<tr>
<td></td>
<td>Maintain liquid intake</td>
<td>Maintain liquid intake</td>
<td>Use aspirator</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Try using a straw</td>
<td>Eat calorie-dense foods</td>
<td>Use medication</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Eliminate caffeine</td>
<td>Increase taste, temperature (colder), and texture sensations of liquids</td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>Use double swallow</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>Learn choking first aid</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>Avoid washing foods down with liquids</td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

ALS, Amyotrophic lateral sclerosis; PEG, percutaneous endoscopic gastrostomy.

(From Yorkston KM, Miller RM, Strand EA: Management of speech and swallowing in degenerative diseases, Tucson, AZ, 1995, Communication Skill Builders.)
Thus dysphagia clinicians should carefully examine any patients presenting with persisting dysphagia for the presence of any related neurologic signs.

**Polymyositis, Scleroderma, and Systemic Lupus Erythematosus**

Polymyositis, scleroderma, and systemic lupus erythematosus are inflammatory muscle diseases more generally classified as connective tissue diseases. A brief but informative summary of dysphagia in these diseases is provided by Sheehan. Polymyositis (dermatomyositis) is an inflammation of striated muscle. It often is initially seen in proximal muscle groups, and when present in the head and neck musculature can contribute to oropharyngeal dysphagia. In these instances clinical characteristics may include nasopharyngeal regurgitation, residue in the pharynx, and airway compromise by food or liquid. Deficits of the cervical esophagus are also frequently reported.

Scleroderma (progressive systemic sclerosis) is an inflammation of smooth muscle tissue. In this respect dysphagia is often esophageal in nature, primarily resulting from dysfunction in the distal third of the esophagus. At some point in the disease process many patients with scleroderma experience solid food dysphagia as a result of esophageal dysfunction. However, oropharyngeal dysphagia also may be seen with this disease.

Systemic lupus erythematosus is a disease process that affects women more frequently than men. The clinical presentation may vary because the disease may involve many organ systems. The time course is also variable. Patients may demonstrate proximal muscle weakness (including head and neck musculature), cranial nerve abnormalities, or deficits in the central nervous system. Often the presentation is of acute deterioration with slow recovery between exacerbations. Many patients report esophageal-based dysphagias.

Other diseases in the category of connective tissue or systemic rheumatic diseases can contribute to dysphagia. The general presentation is fatigue, malaise, pain, reduced appetite, and often dysphagia. Dysphagia may present as oropharyngeal or esophageal or both. Often the determining factor is which muscle groups are involved.

**Muscular Dystrophy**

Muscular dystrophy is another muscle disease that can affect various muscle groups. One type of dystrophy that may directly contribute to dysphagia is oculopharyngeal muscular dystrophy (OPMD). OPMD is a slowly progressive disorder characterized by dysphagia, dysarthria, ptosis, and face and trunk weakness. As the name implies, pharyngeal muscles are likely to be weakened and thus contribute to dysphagia. Depending on the stage of the disease, dysphagia may be mild or severe. Duchenne muscular dystrophy (DMD) is another variant of muscle disorder affecting younger males. DMD is progressive with no known cure. The prevalence of dysphagia in DMD is not well estimated but rates between 18% and 30% have been cited. The pattern of dysphagia in DMD often includes feeding difficulties, but pharyngeal abnormalities have been found more frequently than oral or esophageal difficulties. Dysphagia symptoms are quite varied among individuals with DMD and may include prolonged meal times, difficulty swallowing hard food and thick liquids, and frequent coughing or expectoration during meals.

**Treatment Considerations**

Many diseases that affect lower motor neurons and peripheral muscle groups are progressive and thus present special challenges to the patient and the clinician. As with other neurogenic dysphagias, swallowing interventions often are symptomatic, reacting to the specific set of clinical circumstances presented at any given time. Various strategies may be used; these range from behavioral compensations to diet modifications. The use of strengthening exercises or related strategies may be questionable in some situations. Exercise fatigues muscle groups. If the underlying disease creates weakness in muscles required for swallowing, attempts to over-exercise these same muscle groups may exaggerate the underlying weakness rather than ameliorate it. Available evidence neither supports nor contradicts the use of exercise in progressive neuromuscular disorders. Thus it is important to understand the impact of the underlying neurologic condition on sensorimotor capability of the individual patient.

Clinicians attempting to improve swallowing function also must remember that these patients are receiving ongoing medical care. They often take multiple medications that may be changed from time to time. It is important for the dysphagia specialist to maintain good communication with other members of the health care team to understand better the effects of various medications and make optimum decisions about changes in the dysphagia management plan. Remember, many of these diseases are progressive, necessitating changes in dysphagia management strategies over time. Hillel and Miller provide an excellent perspective on the team approach to management of dysphagia and other bulbar symptoms in patients with ALS. Much of their sage clinical advice is applicable to management of dysphagia in patients with other progressive neuromuscular diseases.

**IDIOPATHIC OR IATROGENIC DISORDERS OF SWALLOWING THAT RESEMBLE NEUROGENIC DYSPHAGIA**

A variety of contributing factors may create a neurogenic dysphagia in the absence of overt neurologic disease. These
factors include undetected vascular deficits (ministrokes),
decompensation with advancing age, decompensation in
complex medical conditions, medication-induced changes,
initial symptoms of a progressive disease, and postsurgical
changes. When dysphagia appears to result from neu-
rologic dysfunction in the absence of overt neurologic
disease or damage, these factors should be considered. A
good rule of thumb is to treat a suspected neurogenic dys-
phagia as the result of a neurologic process until proven
otherwise (see Clinical Corner 3-4).

TAKE HOME NOTES

1. Dysphagia resulting from neurologic disorders reflects
   the underlying sensorimotor characteristics of the neu-
   rologic deficit.
2. Treatment of neurogenic dysphagias is often sympto-
matic but relies heavily on a strong understanding
   of the underlying neurologic process. In many cases
   behavioral treatment interacts significantly with medical
treatment.
3. Many neurogenic dysphagias change over time, neces-
sitating different intervention strategies. Change may
   occur both toward recovery or deterioration of function
   depending on the specific neurologic disease or
   disorder.
4. Medical treatments (including surgery) for various
   neurologic diseases and disorders also contribute to
dysphagia.
5. In the absence of overt neurologic disease, dysphagia
   that appears to be neurogenic should be considered
   reflective of an underlying neurologic cause until proven
   otherwise.

CLINICAL CORNER 3-4: IDIOPATHIC DYSPHAGIA?

A 75-year-old man was referred for evaluation of dyspho-
nia and dysphagia after knee replacement surgery. His
endoscopic swallow examination is presented in Video
3-9 on the Evolve website. Note the nonmoving left true
vocal fold, weakness in the left hemipharynx, and pooled
secretions.

Critical Thinking

1. What factors might contribute to both dysphonia
   and dysphagia in this specific patient?
2. Speculate about the relation between knee surgery
   and dysphonia and dysphagia in this patient.
3. What is the clinical significance of the
   hemiphasaryngeal weakness “on top” of the
   nonmoving left true vocal fold? How might this
   affect treatment planning for this patient?

CLINICAL CASE EXAMPLE 3-1

A 69-year-old man had a brainstem stroke 7 months before
seeking rehabilitation for dysphagia. The patient takes no
food or liquid by mouth and is receiving all nutrition by
PEG. He expectorates saliva into a cup except at nighttime.
Within the past month he has tasted food but not attempted
to swallow. His anxiety level is high about the possibility of
aspiration but he is highly motivated to initiate oral feeding.
He has experienced no chest infections or other complica-
tions since discharge from acute rehabilitation. Clinical
examination revealed a left facial weakness but he was able
to make a strong lip seal. He demonstrated right-body
weakness greater in the arm than the leg, and he was able
to walk with a quad cane. Endoscopic evaluation revealed
slight paresis of the left vocal fold and in the left hemipha-
ryn. Fluoroscopic examination of swallowing function
revealed incomplete swallow attempts with limited hyola-
ryngeal excursion, limited opening of the PES (a small
amount of material entered the esophagus), postswallow
residue for thicker materials, and a small amount of aspira-
tion with thin liquid. He demonstrated a strong reactive
cough to the aspiration and the ability to clear residue back
into the mouth, where it was expectorated.

Interpretation

This patient would be considered in the chronic post-
stroke phase because more than 6 months have elapsed
since his stroke. He has had no swallowing experience
during that period, but the observation that he does not
expectorate at night (and does not complain of a “soggy”
pillow in the morning) possibly suggests that he is swal-
lowing saliva while asleep. The fact that he has tasted food
supports his motivation to undertake aggressive therapy.
His anxiety about aspiration is understandable and may
be a factor to consider once therapy begins. The fact that
he has had no chest infections and no history of trache-
ostomy are positive indications for the respiratory system.
Ambulatory status is considered a positive sign because
active patients are believed to be less susceptible to respi-
atory infections than are bedridden patients. The alter-
nating hemiplegia (left face, pharynx, and vocal fold
versus right side of the body) is characteristic of brain-
stem stroke. The incomplete swallow is characterized by
incoordination and limited excursion of movement of the
hyolaryngeal complex with reduced PES opening. Mate-
rial entering the esophagus is a positive finding, as is the
strong reactive cough and the ability to clear residue.

This patient is a good candidate for direct, intensive
swallowing therapy. An appropriate therapy program for
this individual should address airway protection (either
by choice of material to be swallowed or compensatory
maneuver), hyolaryngeal excursion (increase upward
and forward movement), and swallow coordination (in
some cases slowing the speed of the swallow with pro-
longed maneuvers may accomplish this outcome). If suc-
cessful, the functional outcome should be increased oral
intake of food and liquid.
A 72-year-old woman presented to the clinic with a diagnosis of primary progressive aphasia. The primary complaint was weight loss and unfinished meals. The patient lived independently and attended an adult day-care facility where she reportedly was observed to cough during lunch. Her brother had a history of esophageal disease and a concern was expressed by the family. The patient was ambulatory and presented no overt physical impairments. She was limited in her ability to communicatively interact. Her expressive communication was limited to head nods and a few vocalizations but no meaningful words were produced. She was able to respond appropriately to many basic commands and requests and participated interactively with a dysphagia examination. Oral mechanism examination was unremarkable with no overt signs of corticobulbar deficit. Videofluorographic examination of swallowing was completed. The only mild abnormality was the observation that the patient tilted her head upward as she initiated a swallow and that oral initiation and transit were prolonged. Subsequently, a feeding examination was completed in which the patient was provided a tray of food and liquid (regular-grade diet) and requested to eat. She surveyed the tray of food and promptly began to eat using her fingers. She was handed a fork and used this appropriately until she faced a situation in which she had to cut her food. She was handed a knife and proceeded to use it as a fork. Despite multiple cues she persisted to use the knife as a fork and could not be encouraged to use two tools (knife and fork) simultaneously.

**Interpretation**

This specific case contains features commonly associated with dementias (weight loss, reduced food intake, poor communicative interaction) in addition to a more rare and specific finding, Primary progressive aphasia is a form of dementia in which language skills are impaired early in the course of the dementia, rendering the initial symptoms to those of a progressive aphasia. The observations of utensil use by this patient suggest a form of apraxia that seemed specific to mealtimes and self-feeding. Because at her age and in her situation these social functions were central to her life and her well-being, this form of apraxia had a significant functional impact on her life. The immediate therapy for this individual was environmental. The family was instructed to prepare meals that could be eaten with a single utensil (i.e., fork or spoon). The patient was quite successful with this strategy. Also, it is important to take into consideration the progressive, deteriorating nature of dementia. Although the mealtimes adjustment of a single utensil was effective in the short term, as this disease progressed, this patient would require additional strategies to ensure adequate nutrition and hydration. In this respect, her treatment plan must contain periodic and regular monitoring of the success of any adaptation used to maintain oral food and liquid intake and the nutritional consequences of that intake (see Video 3-3 on the accompanying Evolve website).

**REFERENCES**
