MEDICAL SETTING CONSIDERATIONS

for the

SPEECH-LANGUAGE PATHOLOGIST

Medical Speech-Language Pathology

Series Editors

Kristie A. Spencer, PhD, CCC-SLP

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Series Overview

The Medical Speech-Language Pathology book series provides graduate students, clinicians, and clinical researchers with functional, comprehensive material to enhance practice in a medical setting. The books are designed to bolster transdisciplinary knowledge through infusion of information from neurology, pharmacology, radiology, otolaryngology, and other related disciplines. They capture our current understanding of complex clinical populations, often encountered in medical settings, and offer information to guide evaluation and management strategies. For each clinical population, case studies are used to promote application and integration of the material. Moreover, the handbooks are richly supplemented with figures, tables, and patient samples to enhance accessibility of the information. Each book in the series is authored by experienced professionals and content experts who are able to transform the research literature into clinically applicable and digestible information. The authors integrate theory and practice in a succinct manner, allowing immediate application to everyday practice. This book series advances the medical speech-language pathology community by merging fundamental concepts, clinical strategies, and current theories with research evidence, with the goal of fostering outstanding clinical practice and clinical research.

Preface

The overarching goal of this book is to foster an appreciation of the unique skill set and knowledge base needed by a medical speech-language pathologist (SLP). The chapters contain information applicable to a broad range of medical and rehabilitation settings, while delving deeper into complex topics that merit extra attention, such as neuroimaging methods and medication side effects. Readers will come to appreciate the many roles and responsibilities of the speech-language pathologist in the medical setting, across the continuum of care. They will gain familiarity with broader organizational issues, such as accreditation, billing, and medical team members, as well as the specific clinical populations often encountered in medical settings, such as individuals with Parkinson's disease, delirium, or tongue cancer. Throughout the book, case studies are used to highlight the role of the medical SLP, and to underscore the remarkably dynamic and complex caseloads encountered by the medical SLP.

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patient has transferred to a rehabilitation setting; however, exercises should be considered early, especially in the case of patients who are unable to eat or drink by mouth.

Assessing and facilitating basic communication is important in the acute care setting. Individuals with tracheostomies may benefit from an alternate form of communication, such as writing or pictures, or placement of a speaking valve (Batty, 2009). An individual with dysarthria may benefit from postural adjustments or training in compensatory strategies, such as slowing rate or modifying the speech-breathing pattern (Spencer & Brown., 2018). Alphabet or pacing boards may be beneficial (Yorkston et al., 2010). Use of AAC, such as picture boards, is often effective for patients with aphasia in the controlled hospital environment (Jacobs, Drew, Ogletree, & Pierce, 2004). Family and staff education will be especially important, ensuring that they understand how to set the environment, interact with the patient, and facilitate effective communication.

Direct intervention for impairment level deficits, such as language, memory, and attention, will likely be limited in an acute care setting; however, it is important to be aware of evidence-based methods that may be implemented once the patient is more stable. For example, there is a wealth of literature supporting aphasia treatment, which can be applied to individuals with stroke and TBI. These include methods such as Oral Reading for Language in Aphasia (ORLA; Cherney, 2010), Constraint-Induced Language Therapy (CILT; Pulvermüller et al., 2001), Semantic Feature Analysis (SFA; Efstratiadou, Papathanasiou, Holland, Archonti, & Hilari, 2018), and Verb Network Strengthening Treatment (VneST; Edmonds, 2016), to name a few. Melodic Intonation Therapy (MIT; Sparks, Helm, & Albert, 1974) is sometimes used with patients presenting with co-occurring aphasia and apraxia of speech, though more recent, evidence-based approaches would include treatments such as Sound Production Treatment (SPT; Wambaugh, Wright, & Mauszycki, 2014, 2013). There is positive evidence for interventions targeting many aspects of cognition, including memory, attention, social communication, visuospatial deficits, and executive functions, in patients with TBI and stroke (Cicerone et al., 2011). Training metacognitive strategies is especially important for those with TBI.

NEURODEGENERATIVE DISORDERS

Rene Ruzicka Kanadet

Neurodegenerative diseases are caused by deterioration of neurons in the brain and/or spinal cord. The rate of these diagnoses is expected to increase over the next several decades due to the aging population and increased lifespan (NIH, 2018). At this time, according to the Harvard NeuroDiscovery Center, five million Americans are diagnosed with Alzheimer's disease; one million with Parkinson's disease; 400,000 with multiple sclerosis (MS); 30,000 with amyotrophic lateral sclerosis (ALS, or Lou Gehrig's disease); and 30,000 with Huntington's disease (accessed 2018). While this list is not exhaustive, it captures many of the diagnoses commonly seen by the hospital SLP.

Dementia

Description of the Medical Condition

Dementia is a condition marked by a functional decline in cognitive and/or behavioral symptoms not related to a delirium or major psychiatric disorder (McKhann et al., 2011).

Impairments may include the inability to learn and recall new information; impaired reasoning and judgment; decreased visuospatial abilities, such as difficulty recognizing faces or common objects; impaired language skills; and changes in personality or behavior, such as agitation, social withdrawal, or compulsive behaviors (McKhann et al., 2011). Types of dementia include Alzheimer's dementia, Lewy body dementia, fronto-temporal dementia, and vascular dementia (Smits et al., 2015). The *Diagnostic and Statistical Manual of Mental Disorders* (5th ed.; *DSM–5*; American Psychiatric Association, 2013) now refers to dementia as "major neurocognitive disorder," though the terms continue to be used interchangeably.

Potential Impact on Communication/Swallowing

As described above, cognitive changes are the primary symptoms of dementia, however language impairments increase as the disease progresses. Word finding difficulties are primary in the early stages of dementia, with more noticeable impairments in middle/late stages, characterized by semantic emptiness, and at times becoming nonverbal (Watson, Aizawa, Savundranayagam, & Orange, 2013). Language comprehension also decreases with disease progression. Communication is further impacted negatively by reduced attention, memory, recognition, and behavioral changes. These cognitive-communication impairments negatively affect a person's activities of daily living and, ultimately, their ability to live independently.

Patients with dementia are at increased risk for aspiration. Dysphagia is present in later stage dementia, with diffuse brain lesions impacting the sensorimotor aspects of the swallowing sequence (Rogus-Pulia, Malandraki, Johnson, & Robbins, 2015). Safety during eating is significantly impacted by cognitive impairments in dementia. For example, patients may demonstrate decreased ability to identify, recall, and utilize appropriate utensils during eating; poor attention during mealtimes; decreased ability to self-feed; impulsivity; apathy during meals; and reduced ability to follow instructions. It is suggested that these cognitive impairments may cause difficulty with eating in the early stages of dementia, even before sensorimotor aspects of swallowing have been impacted (Rogus-Pulia et al., 2015). Patients with dementia may also hold/pocket food in their mouths for extended periods of time. Rogus-Pulia and colleagues (2015) summarized swallow study (VFSS and FEES) findings for individuals with dementia, including: longer oral transit times, reduced pharyngeal clearance, and penetration/aspiration in *Alzheimer's disease*; reduced bolus formation and mastication, decreased hyolaryngeal movement, and reduced epiglottic inversion in *vascular dementia*; delayed pharyngeal initiation, pharyngeal residue, and penetration/aspiration in *Lewy body dementia*; and rapid, compulsive eating, large bolus volumes, premature spillage into the pharynx, and pharyngeal residue in *fronto-temporal dementia*. Poor oral care in those with dementia is an additional contributor to aspiration pneumonia risk.

Parkinson's Disease

Description of Medical Condition

Parkinson's disease (PD) is a progressive movement disorder. It is characterized by tremor at rest, rigidity, bradykinesia, postural instability, flexed posture, and freezing (Jankovic, 2008). Additional nonmotor symptoms often include changes in sleep, smell, and cognition. Age of onset is generally between 55 and 75 years. Severity of symptoms may be rated by the Hoehn and Yahr Scale (Goetz et al., 2007; Hoehn & Yahr, 1967), which ranges from

stage 0 (no signs of disease) to 5 (wheelchair bound or bedridden unless assisted), or the Unified Parkinson's Disease Rating Scale (UPDRS; Goetz et al., 2007), a comprehensive rating system used to track the motor and nonmotor characteristics of PD. Motor symptoms are attributed to degeneration of dopamine-producing neurons within the substantia nigra pars compacta. Presence of a-synuclein, a substance in Lewy bodies, is also indicated in PD (Exner, Lutz, Haass, & Winklhofer, 2012). PD is treated with dopamine-replacement medications, such as Levodopa, to reduce severity of symptoms. Deep brain stimulation (DBS) to the globus pallidus or subthalamic nucleus is also used, either in combination with Levodopa or in an effort to reduce drug therapy due to intolerance (St. George, Nutt, Burchiel, & Horiak, 2010).

Potential Impact on Communication/Swallowing

Hypokinetic dysarthria is typically associated with PD. Distinctive characteristics include reduced vocal intensity, monopitch, monoloudness, decreased articulatory precision, and accelerated rushes of speech (Duffy, 2013). Palilalia, or compulsive repetition of words or phrases, may also occur in patients with PD. Cognitive-communication impairments are common, with cognitive deficits ranging from mild cognitive impairment (MCI) to dementia (Litvan et al., 2012). Early cognitive impairments are reported in executive function, attention, visuospatial skills, and memory, notably retrieval skills (Nazem et al., 2009). Mild cognitive impairment is reportedly present in anywhere from 35% to 42.5% of patients at the time of diagnosis, and 50% will be diagnosed with dementia within 10 years (Cosgrove, Alty, & Jamieson, 2015).

Dysphagia is a common condition associated with PD, occurring in approximately 80% of individuals with PD (Kalf, de Swart, Bloem, & Munnecke, 2012). Reported oral phase deficits include reduced bolus formation and mastication, delayed initiation, tongue pumping, premature spillage into the pharynx, piecemeal deglutition, and oral residue. Pharyngeal phase deficits include delayed laryngeal movement, reduced epiglottic inversion, and decreased pharyngeal constriction, resulting in pharyngeal residue and increased rates of penetration and aspiration (Michou, Baijens, Rofes, Cartgena, & Clave, 2013). Lingual weakness may also be implicated in dysphagia (Pitts, Morales, & Stierwalt, 2018).

Multiple Sclerosis

Description of Medical Condition

Multiple sclerosis (MS) is a chronic, inflammatory, autoimmune disease of the central nervous system and is cited as one of the most common causes of neurological disability in young adults (NIH, 2018). The initial symptoms of MS typically occur between 20 and 50 years of age, and women are approximately 3 times more likely to develop MS compared with men (Gooch, Pracht, & Borenstein, 2017). Interestingly, the prevalence of MS varies by geographic location, and generally increases the further one travels from the equator (Simpson, Blizzard, Otahal, Van der Mei, & Taylor, 2011). Reasons for this geographic influence are not entirely clear, but may be linked to vitamin D deficiency or genetic differences.

Multiple sclerosis is marked by damage to the myelin sheath of neurons in the brain and spinal cord with clinical manifestations dependent upon what neurons have been affected. These may include changes in movement, sensory perception, cognitive function, speech, and swallowing. Differential diagnosis is often difficult with MS due to similar presentation with diseases including Lyme disease,

lupus, and paraneoplastic disease (McDonald et al., 2001). There are four types of MS with relapse-remitting being the most common. This type of MS is characterized by exacerbation periods followed by recovery/remission, though full recovery between relapses may not occur, and progressive decline is seen over time. Additional types of MS include primary-progressive, characterized by progressive deterioration from onset; secondary-progressive, with initial relapsing-remitting followed by progression; and progressive-relapsing multiple sclerosis, characterized by acute relapses but with continuing progression between relapses (Miller & Leary, 2007).

Potential Impact on Communication/Swallowing

Dysphagia, dysarthria, and cognitive-communication impairments are possible in patients with MS. The clinical presentation is dependent upon what areas of the brain have been affected by demyelination and neurodegeneration. Dysarthria can be any type, with spastic-ataxic being the most commonly cited mixed dysarthria (Duffy, 2013). Speech characteristics associated with MS include reduced volume control; harsh voice; imprecise articulation; impaired stress patterns, rate, and breath support; and variable pitch (Darley, Brown, & Goldstein, 1972; Hartelius, Theodoros, Cahill, & Lillvik, 2003).

Cognitive changes may occur early, preceding physical impairments, and can occur with any type of MS (Patti, 2009), though it is highest in the secondary progressive population (Trenova et al., 2016). Presentation is heterogeneous, though impairments are most commonly seen in memory, attention, processing speed, visuospatial skills, and executive functions. Fatigue and depression may also contribute to cognitive-communication deficits (Patti, 2009; Trenova et al., 2016).

Dysphagia is reported in anywhere from 17% to 65% of patients with MS (Giusti & Giambuzzi, 2008; Tassorelli et al., 2008). Characteristics of dysphagia include impairments in the oral phase due to lingual, labial, and/or velar involvement; premature spillage of material into the pharynx and/or larynx; impaired tongue base retraction; decreased pharyngeal contraction; reduced laryngeal closure; and decreased pharyngeal and/or laryngeal sensation (Prosiegel, Schelling, & Wagner-Sonntag, 2004; Tassorelli et al., 2008). These impairments result in decreased oral and pharyngeal clearance, penetration/aspiration, and apnea during or after eating (Giusti & Giambuzzi, 2008; Tassorelli et al., 2008). Some patients with MS also have decreased relaxation of the upper esophageal sphincter, which contributes to pharyngeal residue (Giusti & Giambuzzi, 2008; Prosiegel, Schelling, & Wagner-Sonntag, 2004; Tassorelli et al., 2008).

Amyotrophic Lateral Sclerosis

Description of Medical Condition

Amyotrophic lateral sclerosis (ALS) is a mixed upper and lower motor neuron disease characterized by degeneration of motor neurons in the brain and spinal cord. Typical age of onset is mid-to-late 50s, and death usually occurs within 3 to 5 years (Brown & AlChalabi, 2017). Most cases of ALS begin with limb weakness, eventually spreading to most muscles, including those of respiration. Approximately one-third of cases, however, are bulbar, which is characterized by earlier difficulty speaking and swallowing. Pseudobulbar affect may be seen. Some patients with ALS will also present with fronto-temporal dementia. Additional types of motor neuron disease include progressive muscle atrophy (PMA), primary lateral sclerosis, and progressive bulbar palsy

(PBP), though ALS represents approximately 80% of total cases (Chieia, Oliveira, Silva, & Gabbai, 2010).

Potential Impact on Communication/Swallowing

Dysarthria and dysphagia occur in all patients with ALS, though persons with bulbar onset will experience severe speech and swallowing symptoms earlier in the disease progression. The typical dysarthria associated with ALS is mixed spastic-flaccid due to involvement of the upper and lower motor neurons (Duffy, 2013). This dysarthria commonly manifests as slow rate of speech, imprecise articulation, hypernasality, monopitch, and harsh or breathy vocal quality. As the disease progresses, 80% to 95% of people with ALS will become unable to use natural speech for functional communication, and most will lose their ability to speak (Beukelman, Fager, & Nordness, 2011).

Dysphagia is one of the earliest symptoms in individuals with ALS with a bulbar onset, though it occurs in all patients with ALS and typically leads to percutaneous endoscopic gastrostomy (PEG) placement for enteral nutrition. Dysphagia symptoms include chewing fatigue, drooling, nasopharyngeal reflux, coughing and choking, impaired bolus formation, and difficulty with oral transit (Hadjikoutis & Wiles, 2001). Early dysphagia in ALS patients was described by Teissman et al. (2011) as primarily pharyngeal, characterized by weak pharyngeal constriction and residue in the valleculae and pyriforms, whereas impaired bolus preparation and transit was evident in more severely impaired patients. Atypical respiratory patterns are also present during swallowing with inspiration after the swallow, longer apnea during the swallow, and multiple swallows per bolus (Hadjikoutis & Wiles, 2001).

Cognitive deficits are evident in approximately 30% of patients with ALS and are characterized by impairments in fluency, language, social cognition, verbal memory, and executive functions (Beeldman et al., 2016). It is estimated that 15% to 20% of ALS patients will present with fronto-temporal dementia (Abrahams, Newton, Niven, Foley, & Bak, 2014; Brown & Al-Chalabi, 2017); the co-occurrence of these disorders has been linked to genetic mutations (Turner et al., 2017). Frontotemporal dementia (FTD) is marked by behavioral and personality changes.

Huntington's Disease

Description of Medical Condition

Huntington's disease (HD) is an autosomal-dominant genetic disorder. The primary motor characteristic is chorea, which is characterized by involuntary jerking and writhing movements. Other common symptoms include dystonia, incoordination, slow saccadic eye movement, cognitive decline, motor impersistence, and behavioral changes (O'Walker, 2007). The age of onset for HD is typically middle-age though a juvenile onset is possible between the ages of 2 and 20. Genetic testing can be completed to confirm HD even before symptoms present. Although there is no treatment to slow disease progression, management strategies may include pharmacological treatment of chorea and pharmacological and nonpharmacological treatment of psychiatric and behavioral disturbances (Frank, 2014).

Potential Impact on Communication/Swallowing

Cognitive impairments are a hallmark characteristic of HD and impact participation in daily living tasks and communication situations.

Impairments are seen in attention; memory; visuospatial processing; executive functions, including problem solving, thought flexibility, and planning; and emotional processing, including interpretation of emotions through facial expression and voice (Montoya, Price, Menear, & Lepage, 2006). These deficits become severe in later stages of the disease.

Speech intelligibility is impacted by choreic movements, resulting in a hyperkinetic dysarthria. The severity of the person's dysarthria is related to the severity of the chorea. Movements of the jaw, tongue, and lips can affect articulation. Irregular articulatory breakdowns, prolonged phonemes, and abnormal lengthening of vowels are characteristic (Duffy, 2013). Unpredictable movements can also result in changes in respiration, such as forced and involuntary inspiration or expiration; volume, such as excess loudness; and vocal quality, including a strained vocal quality or voice arrest.

Dysphagia is likely to occur as HD progresses (O'Walker, 2007). Characteristics of dysphagia with chorea include rapid lingual movements, decreased coordination of the swallow, prolonged laryngeal elevation, and poor coordination of respiration and swallowing (Kagel & Leopold, 1992). Kagel and Leopold (1992) further described that bradykinesia in patients with HD can lead to mandibular rigidity, slow lingual chorea, and coughing on foods and liquids. Pharyngeal retention and aspiration were identified through videofluoroscopy in this same study. Self-feeding is also an issue for patients with HD (Bilney, Morris, & Perry, 2003). Involuntary movements may limit the ability to bring food and utensils to the mouth in a controlled manner, causing reduced oral acceptance and control and/or anterior loss. Body movements may also increase aspiration risk. In a retrospective study by Heemskirk and Roos (2012), 55% of the reviewed patients died of pneumonia, and a majority of these patients could be further identified as having aspiration pneumonia.

Role of the SLP in Neurodegenerative Disorders

The role of the SLP changes with disease progression and may start before there has even been a confirmed diagnosis. A thorough oral mechanism exam and evaluation of speech and swallowing may offer neurologists and other medical providers valuable information during the diagnostic process. These evaluations, in addition to assessment of cognitive-communication, should be completed at all phases of disease progression.

At early stages of disease, an important role of the SLP is education, especially when there are no overt symptoms of dysphagia, dysarthria, or cognitive-communication deficits. It is known that most persons with neurodegenerative diagnoses will experience these impairments, as described above; therefore, it is important to consider educating the patient and family on what to expect, when to seek medical attention, and the role of the SLP throughout the progression of their disease. Education on clinical signs of aspiration and other warning signs for dysphagia, such as weight loss, fevers, and pneumonia is especially important. It is also important that the SLP complete assessments in these early stages. Subtle signs of dysphagia, dysarthria, and cognitive-communication changes may not be reported. Underreporting of symptoms may occur secondary to progressive adaptation to slow changes. Assessments are warranted to identify impairments and encourage early intervention and/or to serve as a baseline for future assessments.

Continued assessment is important at all phases of disease progression in order to provide updated recommendations, especially

with regards to dysphagia, as aspiration pneumonia is a leading cause of death in these populations (Walshe, 2014). Patients with previous knowledge of their dysphagia may be resistant to further diagnostics or feel that they are already well informed. Provide education and encouragement. Diet modifications and compensatory strategy training are important as neurodegeneration progresses. Cognitive abilities should always be taken into consideration when making recommendations, and family or caregiver training is crucial.

When the progression of the disease has become very severe, advanced diagnostics, such as VFSS, may not be appropriate. A discussion with the patient's family to determine priorities and end-of-life wishes should occur prior to completing these tests. Quality of life must always be considered. It is imperative that the SLP provide thorough education on dysphagia and the risks of an unrestricted diet; however, the patient/family's decision must always be respected. Including the patient's physician on the conversation is recommended when discussing diet liberalization.

During an acute hospitalization, patients with neurodegenerative diseases will likely be encountered with a different admitting diagnosis. For example, a patient with dementia may be admitted for pneumonia, or a patient with PD may be admitted following a fall and subsequent hip fracture. In these cases, the SLP may not be consulted or may be consulted for an isolated assessment, such as swallowing. During short admissions, or when a patient is acutely ill, treatment of speech, swallowing, and/or cognitive-communication skills may not be feasible or a priority; however, referral to rehabilitation services should be encouraged. Initial education to patients and families on the rationale for intervention may increase understanding and follow-through post-discharge from the acute care setting. SLP recommendations may also contribute to physician decision making regarding next level of care, for example, if considering discharge home or to inpatient rehabilitation.

Although not curative, there is positive evidence for rehabilitation with neurodegenerative populations. Lee Silverman Voice Treatment (LSVT) is a well-known example of treatment for patients with dysarthria related to PD (Sapir, Spielman, Ramig, Story, & Fox, 2007). Use of expiratory muscle strength training (EMST) has shown improvements in maximum expiratory pressure and hyolaryngeal elevation in ALS (Plowman et al., 2016) and PD (Troche et al., 2010), as well as improving cough strength in patients with moderate MS (Chiara, Martin, Davenport, & Bolser, 2006). Cognitive-communication treatment may include strategy training and use of repeated cognitive exercises resulting in a training effect (Lovera & Kovner, 2012). Spaced Retrieval Training (SRT) has shown success in increasing new learning in patients with dementia (Hopper et al., 2013; Oren, Willerton, & Small, 2014), as well as use of SRT paired with external aids for compliance with swallow strategies (Benigas & Bourgeois, 2016). Alternative and augmentative communication (AAC) should be considered as a supplement or alternative to verbal communication for patients with dysarthria. This may be low-tech, such as use of an alphabet board or writing, or a high-tech speech generating device (Fried-Oken, Mooney, & Peters, 2015). Numerous access methods for speech generating devices are available for individuals with pronounced physical limitations, such as eye gaze, head movement, or foot movement. Providing and training use of an AAC device is recommended early in the progression of diseases, such as ALS, and is associated with increased quality of life (Körner et al., 2013).

Specific diagnosis, stage of progression, previous SLP assessment and treatment, and patient/family wishes must all be considered