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1 Purpose and Scope

This is a clinical book about swallowing. Swallowing is the complex activity of suspending respiration while moving saliva, food, drink, or any other substance from the mouth through the pharynx and esophagus into the stomach. Traditionally, the nonrespiratory portion of this complex process is divided into oropharyngeal and esophageal stages. An appalling number of congenital and acquired conditions can disrupt the process, resulting in what are commonly referred to as either *oropharyngeal* or *esophageal* dysphagias. This book is dedicated to *oropharyngeal dysphagia in adults resulting from movement disorders*.

This book is intended for the clinician and for clinical use. It is written to support understanding, evaluation, and behavioral treatment of the swallowing problems resulting from the full range of movement disorders, such as Parkinson's disease (PD), multiple system atrophy (MSA), Huntington's disease (HD), and dystonia. Our wish is that if a patient with even a rare movement disorder appears in the clinic, this book will offer some guidance.

Separate chapters are devoted to specific disorders such as PD or to classes of movement disorder such as ataxia and dystonia. In these general chapters such as the entry on dystonia, individual conditions such as Meige syndrome will

appear alongside other conditions. This organization was chosen because so little is known about swallowing in most of the disorders characterized by movement abnormality. PD is an obvious exception; hence, this condition has its own chapter. Even the general chapters differ in length, depending on what is known. These chapters have the same organization, including definitions, disorders, epidemiology, general and swallowing signs and symptoms, and special evaluative and treatment considerations. The evaluation and management options are based on the evidence, when it exists, our own clinical experiences, and the realities of the modern health care environment.

The book begins with short chapters on definitions; anatomy and physiology of the normal swallow; the history and chart review; and the clinical, videofluoroscopic, and videoendoscopic swallowing examinations. These chapters are followed by entries outlining general and specific principles governing behavioral management and discussions of behavioral treatments divided, with some trepidation, into rehabilitative and compensatory approaches. The disorder chapters comprise the book's final section. The book ends with appendices of particularly useful information.

This book is meant for the practitioner. Where clinical arguments about

2 DYSPHAGIA IN MOVEMENT DISORDERS

what to expect or do exist, we take a stand. Where nothing is known for sure, we take a chance. These are the same stands and chances we take in our own clinical practices.

2 Definitions and Classifications

Normal swallowing and dysphagia are defined in this chapter. The stages of swallowing, a traditional conceit that fits only uncomfortably with the act of moving food and drink from the mouth to the stomach, are also described briefly. In some instances a movement disorder may disproportionately affect one stage or another; therefore, we have preserved the idea of stages. Clinicians are asked to recognize, however, that the majority of swallowing problems affect more than one stage and that these divisions are somewhat arbitrary (Martin-Harris, Michel, & Castell, 2005). The second section of this chapter is devoted to a discussion of definition and classification in movement disorders. The problems of classification of these disorders have never been solved to everyone's satisfaction. Certainly, our solution will not be universally applauded. It works for us clinically so we have used it here.

NORMAL SWALLOWING

Normal swallowing is the complex cognitive, sensorimotor act of moving any bolus from the mouth to the stomach. Normal swallows are *safe, efficient, and satisfying*.

Stages of Swallowing

Swallowing, traditionally, is divided into four stages: oral preparatory, oral, pharyngeal, and esophageal. The *oral preparatory stage* is highly volitional and characterized by chewing and mixing the bolus with saliva. The *oral stage* is characterized by the final formation of the bolus into a convenient shape and posterior movement of that bolus through the faucial pillars and into the pharynx to trigger the pharyngeal stage of swallowing. For our purposes, we generally combine the oral preparatory stage and the oral stage and have done so throughout this book. In the *pharyngeal stage*, the bolus is moved through the pharynx and rostral esophagus. This striated muscle section of the esophagus is called the upper esophageal sphincter (UES). The *esophageal stage* involves movement of the bolus through the esophagus into the stomach. It is useful to think of these stages as overlapping and interacting in what can be seen as a complex, sequential response. It is also common to view the stages as existing along a continuum of automaticity, with the esophageal stage being most automatic and the oral preparatory stage the least.

DYSPHAGIA

Dysphagia is defined as *disordered movement of the bolus from mouth to stomach due to abnormalities in the structures critical to swallowing or in their movements*. Dysphagia involving any one or combination of the stages can make swallowing unsafe and put a patient at risk for malnutrition, dehydration, fatal choking episodes, and pulmonary consequences of aspiration. It can make the swallow inefficient and require that the patient take hours to eat. It can make eating unpleasant because the patient fears choking or grows weary of dietary restrictions.

Normal Movement

Normal movement depends minimally on *posture, strength, timing, tone, steadiness, and praxis or skill*.

Movement Disorders

Classic neurology identifies a set of movement disorders, although all textbooks and practitioners do not agree on all the appropriate members of this set. Fernandez, Rodriguez, Skidmore, and Okun (2007) begin by categorizing all movement abnormalities as either hyperkinetic, characterized by an excess of movement, or hypokinetic, characterized by a “paucity” of movement. For these authors, the two main *hypokinetic* abnormalities are what they call *parkinsonism* and *rigidity*, whereas the *hyperkinetic* abnormalities are *chorea*, *dystonia*, *myoclonus*, *tic*, and *tremor*. Restless leg syndrome is also included as a major component of

the hyperkinetic conditions, but no further mention of it will occur in this book on swallowing. Experienced clinicians recognize that other often-identified movement abnormalities such as athetosis and ballismus are absent from the list. Fernandez and colleagues (2007) include these two with chorea and opine that these three abnormalities may actually exist on a continuum.

It is unclear how most of these conditions affect swallowing and if their effects are unique. Clinical experience suggests that some of these abnormalities (e.g., tic) have less frequent and disastrous effects on swallowing than do others (such as Parkinson’s disease [PD]). For all, the severity of the abnormality may determine the swallowing effect, as may the coexistence of other abnormalities such as weakness.

Modern clinical neurology usually begins with an emphasis on the disease state (i.e., disorder) such as PD or multiple system atrophy (MSA) rather than movement abnormalities per se. The movement abnormalities are used primarily in support of the movement disorder diagnosis. Logic would seem to dictate individual chapters on each of the movement disorders. Unfortunately, that approach was unworkable. More than one hundred movement disorders have been identified. Some, such as PD, are frequent and the resulting dysphagia has been widely studied. Others, such as MSA, are more rarely encountered and dysphagia is less well studied. The result would have been an inordinate number of chapters of wildly varying lengths and specificity.

Equally unworkable would have been chapters devoted to each of the movement abnormalities. The rarity of some is but one complication. In addition, some

such as tic often have no influence on swallowing. More critical is that some conditions that are traditionally regarded as movement disorders (such as PD) may present with most of the abnormal movements listed. Likewise, some abnormalities such as tremor may be present in a majority of the commonly recognized disease states.

Therefore, the chapter headings in this volume are a mixture of diseases (e.g., PD, Wilson's disease [WD]) and of move-

ment abnormalities (chorea, dystonia, dyskinesia). Given this format, the Index will often serve as a better guide to the content than will the Table of Contents.

As is obvious from the foregoing, movement disorders present as many challenges for the writer as for the clinician. We try to counter that difficulty a bit by being as specific about how to identify and manage the swallowing disorders of those with movement disorders as our experience and the data allow.

5 Clinical Swallow Examination

The clinical swallow examination (CSE) is the heart of clinical practice. Attempts to relegate it to the status of screening examination are to be resisted. This examination sets the agenda for all else that happens between the patient and clinician. Its competent performance is the measure of a clinician's excellence.

EXPECTATIONS OF THE EXAMINATION

The CSE is the sophisticated clinician's most powerful tool. A maximum expectation of the procedure is that it will inform the perspicacious diagnostician about:

1. The likelihood that a person has a dysphagia
2. Selected signs, especially drooling, loss of bolus between the lips, reduced chewing, and even rocking of the bolus
3. The likelihood of penetration and aspiration with signs like wet voice, coughing, choking, and changes in breathing
4. Estimate of velopharyngeal function, realizing that velopharyngeal function in swallow will differ from that

function in speech or upon observation of elevation during /a/

5. Estimate of laryngeal function with the same caveats
6. Determination of peripheral upper extremity mobility
7. Determination of cognitive status, including memory and attention
8. Insight into the patient's ability to perceive and manipulate food and drink and reaction to food and drink
9. With the right clinical manipulations, determination of what treatments are most likely to be effective
10. Other problems patient may have, such as poor dentition, that could influence evaluation or treatment
11. Whether or not an instrumental examination is necessary for further assessment

Caveat

Few, however, are the clinicians willing to grant this much power to the CSE. Typical complaints are that aspiration may be missed and that a physiologically oriented treatment cannot be derived. A clinical book is not the forum for a discussion of these issues. Rather, it is our position that in the right hands, the CSE is

9 Classification of Treatments

Treatments can generally be divided into compensatory and rehabilitative approaches, although the boundary of these two is indistinct and some methods may be more one or the other or neither, depending on how they are applied. We have preserved the distinction because we think it focuses clinicians on a major issue in dysphagia management—*the possibility that too many clinicians limit themselves to compensatory approaches*. In our view, rehabilitation should be the primary aim of treatment, and compensatory methods should be part of the plan only in conjunction with rehabilitation or if rehabilitation fails or is impossible. For us the reason is simple. The brain learns to do what it is forced to do. A brain whose body is tube fed will devote its critical real estate to activities the body is still performing.

Compensatory techniques are aimed at an immediate influence on safety and adequacy of nutrition and hydration. They are applied when a person needs immediate help either in the absence of rehabilitative efforts or in addition to them, or when a person cannot be expected to complete or profit from rehabilitative efforts. Once a decision in favor of compensatory approaches is made, it remains to choose the right ones and to derive a sense of security that the approach will indeed support safe, adequate nutrition

(pleasurable is sometimes possible as well). These are not easy decisions, although it is our experience that many clinicians seem to behave as if they can be made almost reflexively. Safety and adequacy are difficult to judge because evidence for both evolves over time. It may take days or weeks for malnutrition or dehydration to emerge, and the same is true for respiratory illness related to aspiration of food or drink. Thus, the clinician is left to predict based on the results of clinical and instrumental examinations, history, and clinical judgment. Chapter 13 describes the major compensatory treatments available to the dysphagia clinician working with patients with movement disorders (and often other conditions as well).

Rehabilitative techniques are designed and applied to improve swallowing function. They do so by improving the skilled movements necessary to a safe, efficient, and satisfying swallow or by altering the assumed pathophysiology underlying the abnormal swallow. For example, if weakness or reduced endurance were posited to be the reason for swallowing signs in an individual patient, then a rehabilitative technique is one that improves strength or endurance. This approach is in contrast to a compensation approach in which impaired skill, strength, or endurance would simply

be accommodated by postural change, diet modification, or some other alteration in how or what the patient is eating and drinking. In addition to strength or endurance, rehabilitative techniques involve efforts to change sensory thresholds, tone, timing, or coordination. Chapter 14 contains descriptions of the rehabilitative procedures potentially useful in movement disorders. The challenges of evaluating rehabilitative treatment effects are similar to those for the compensatory methods. However, the clinician faces other challenges as well. The foremost of these is that rehabilitation usually requires more of the clinician's and patient's time than do compensatory methods, and the effects are often delayed by weeks. As but one example, it may take

6 or more weeks for strength to improve, despite nearly daily exercise.

A final challenge in executing intelligent treatments, especially rehabilitative ones, is that traditional training often ignores the principles that guide such treatments. Students are taught how to do the techniques but not how to select them or how to evaluate new techniques when they appear, often to great clinical acclaim. Therefore, the next two chapters (Chapter 10 and Chapter 11) offer brief lists of principles for both compensatory and rehabilitative approaches. Once learned, they seldom need to be reconsulted, but the rewards of reading these chapters carefully may be substantial. It may prevent use of ineffective or otherwise inappropriate methodologies.

10 Principles of Compensatory Treatments

Compensatory as used in this chapter refers to approaches that try to accommodate each person's dysphagia rather than trying to improve it. Three broad classes of activity traditionally are defined as compensatory:

1. *The first is change in posture.* The usual ones described in the treatment literature are the *head turn* and *chin tuck*. The goal of such adjustments is to put a patient in a posture that allows safer, more efficient passage of the bolus through the mouth and pharynx and into the esophagus.
2. *The second is change in the food and/or liquid preparation.* The usual changes are in food consistency—regular, mechanical, soft, pureed—and in the viscosity of the liquid using thickeners.
3. *The third is change in approach to eating.* Any number of manipulations fit here. Common sense changes are bolus size, rate of eating or drinking, and bolus order, such as when liquids and solids are alternated.

To the casual practitioner making these changes may seem simple enough, so simple that principles should not be necessary. We believe, however, that deci-

sions about compensatory approaches are (or should be) difficult. These difficult decisions may be improved if made with a few principles in mind.

PRINCIPLE 1

The classification of all swallowing treatments into rehabilitative and compensatory is based on tradition and not on data. Therefore, the classification is likely to be wrong. Consider the use of thickened liquids. It is assumed that their use is compensatory. The patient, for example, who aspirates thin liquids and is placed on thickened liquids may well be safer. It is possible he may also swallow more than if placed NPO (*nil per os* or, more commonly, nothing per oral). Does increased swallowing translate into improved swallowing? The answer to this question remains unknown, but we suspect continuing to swallow food and drink is more beneficial than remaining NPO and reducing the number of swallows completed. This may be especially true if the clinician couples an altered diet with instruction that the swallows be quicker, more complete, or in some other way more skillful.

PRINCIPLE 2

Despite principle 1, *we advocate using rehabilitative treatments before or in consort with compensatory ones as a first order of clinical business.* For example, in the case of a patient needing thickened liquids because of aspiration through an inadequate laryngeal valve, a rehabilitation program to increase laryngeal closure should be introduced if the patient has even a modest chance of understanding and completing it.

PRINCIPLE 3

Not all compensations, even if they appear effective on the videofluoroscopic swallow examination (VFSE), will keep a patient safe from the effects of aspiration. The rule is that aspirators aspirate. Additionally, as noted by Logemann and colleagues (2008), the effect of aspiration of thickened liquids remains unknown and may increase the likelihood of pulmonary complications.

PRINCIPLE 4

Compensatory techniques, especially if they involve postural adjustments, may be unacceptable to some patients. In this case, the clinician is wise to select one or more rehabilitative techniques exclusively or do nothing.

PRINCIPLE 5

If selected, the planned duration of compensatory techniques should be considered and that duration explained to the patient prior to introduction. If a compensatory technique is not rehabilitative, at least for an individual patient, and is therefore likely to be part of the patient's swallowing for a lifetime, the patient should know that. Few of us would choose to tuck our chins for years on end.

PRINCIPLE 6

Combinations of compensatory techniques or of compensatory and rehabilitative techniques are likely to have the greatest positive effect. For example, a chin tuck and head turn combined with thickened liquids and a laryngeal closing technique may constitute a total therapy for an individual.

PRINCIPLE 7

Lack of patient compliance with compensatory methods is likely to be high. Evaluating the acceptability of any suggestions about compensations and avoiding their use when a patient is clearly unresponsive may save dollars, time, and travail.

11 Principles of Rehabilitative Techniques

Rehabilitative techniques are of two basic types, depending on target. *Type one techniques* aim to change the underlying pathophysiology, principally weakness and reduced endurance. *Type two techniques* aim to increase skill. To achieve either aim, treatments must be guided by one or more of a limited number of principles. These principles derive from what is known about how to influence *plasticity*, or a person's ability to change with experience. We identify three kinds of plasticity: *muscle*, *behavioral*, and *neural*. This chapter contains a description of principles that guide efforts to influence one or more of these three kinds of plasticity. We felt compelled to review them because treatments independent of such principles are likely to be useless at best and dangerous at worst. All methods do not conform to all principles, nor should they. Indeed, methods may seem to fit some and violate others. In this latter incidence, it is up to the clinician to make choices about how to proceed. Please refer to Chapter 14 for more details on the rehabilitative methods discussed below.

MUSCLE PLASTICITY

Muscle plasticity as used in this volume, refers to *chemical, cellular, and muscle*

fiber changes secondary to appropriate training to increase strength and endurance. Two main principles govern treatment to improve strength or endurance.

The first principle is *specificity*. In its simplest form, specificity refers to the phenomenon that persons improve the specific behaviors they practice. The implication for swallowing is that *treatment targets should ideally include components of the behavior to be improved, swallowing in this case.* According to this criterion, tongue protrusion and lateralization against the resistance of a tongue blade fail to meet the specificity principle, as swallowing does not require these two movements. The hard swallow, on the other hand, does meet the criterion. Lee Silverman Voice Therapy (LSVT) approximates it in at least one way: it requires, as does swallowing, laryngeal adduction. Tongue strengthening using the Iowa Oral Performance Instrument (IOPPI) (Robin, Goel, Somodi, & Luschei, 1992; Robin, Somodi, & Luschei, 1991) also satisfies this criterion, as strengthening of tongue tip and tongue back elevation is the goal. Using a method that fails to meet the specificity principle is not necessarily to be avoided. On the other hand, influence of methods that violate the principle may be slow to work and their effects minimal or even nonexistent. Clinicians need to decide if they can

risk these outcomes with methods failing to meet the specificity criterion. In favor of many such methods, however, is that they may conform to another principle, that of overload.

The second principle, *overload*, guides all procedures whose aim is strengthening or increasing endurance. Overload means *the treatment requires more strength or endurance than is necessary for the normal performance of that task*. Simply swallowing, unless it is against resistance or in some other way made more difficult, fails to meet the overload principle. Therefore, the classic notion that swallowing is the best treatment for swallowing needs to be critically evaluated. On the other hand, the Masako maneuver, during which the person swallows with his tongue protruded, conforms to the principle because successfully swallowing in this manner is very difficult. Similarly, expiratory muscle strength training (EMST) meets this criterion by requiring the person to exhale against systematically increasing resistance.

Other principles are assumed within the overload principle. Selected ones of these are worth reviewing, despite our near total ignorance about how they apply in swallowing rehabilitation.

One of these concerns is the *amount of resistance* to be added to the performance for strength to increase. The guideline borrowed from the physical training literature is that training should occur at 60–75% of maximum strength (Powers & Howley, 2003). Most of the traditional strength training procedures, such as using tongue blades to resist tongue protrusion as one example, are seldom applied in light of this principle. Newer methods, such as respiratory muscle strength training or tongue strengthening with the IOPI, however, can be. And

it may even be that strengthening with a tongue blade can be made to meet this criterion at least roughly, and roughly may be sufficient (Lazarus, Logemann, Huang, & Rademaker, 2003).

A second principle is *intensity*. At a minimum, rehabilitative treatments, especially those to improve strength or endurance, should be conducted for 6 weeks. Longer is almost always better. Similarly, a minimum number of sessions per week is three, and six is preferable, though most need not be completed in the clinic. Finally, the number of repetitions per day has never been established in the swallowing literature, but our rule of thumb is a minimum of 25 per day for strengthening and 100 per day (at a lower load) for endurance. Clearly, these are rigorous requirements and much beyond the clinician's time or third-party payer resources. Hence the absolute necessity of a home therapy program accompanied by a diary, so that each patient can record the number of repetitions per day, number of days per week, and number of weeks over which treatment is extended. Patients must of course return to the clinic regularly, so that gains can be measured and new targets, such as amount of resistance, established.

A third is the *necessity for continuing exercise* if maintenance is to be established. Strength and endurance decline when training stops. Therefore, maintenance programs are a necessity. The intensity of these can be reduced and clinical follow-up is often unnecessary. This principle can be discouraging for patients. Many are not compliant with this notion and some will refuse to enter into a treatment once they know the obligation. It is a form of clinical dishonesty, however, to initiate certain programs without mentioning the maintenance principle.

14 Rehabilitation Techniques

This chapter summarizes rehabilitative techniques. Those rehabilitation techniques requiring special instrumentation or special certification are briefly described and references included so the interested reader can consult primary sources. Those that require clinician knowledge and a minimum of instrumentation are described more completely. Admittedly, a bit of magic goes into the division of rehabilitative and compensatory. Traditionally, and in this volume, changes in food texture or fluid viscosity are not included as rehabilitation techniques. However, there is no evidence at all that some such changes are devoid of rehabilitative function. Indeed, we can imagine conditions in which a clinician might impose an increasingly demanding swallowing adequacy requirement on thick liquid swallows. In this case, thickened liquids might earn a place in the list of rehabilitative techniques. A clinical book, however, is not the battleground for fighting classification battles. Therefore, our goal with the division into rehabilitative and compensatory is to classify them traditionally and by central tendencies. No division, including this one, is absolute, and certainly revisions in how methods are to be classified will occur. For now, however, here is how it seems to us.

LEE SILVERMAN VOICE THERAPY (LSVT)

Description of Method

This method of maximum performance training, which when introduced for the treatment of speech in men and women with Parkinson's disease (PD), emphasized the concept of "Think Loud," has now been introduced into swallowing treatment (El Sharkawi et al., 2002). Administering the treatment requires special training and certification. As a result, the method will be described only in brief detail here. Readers are cautioned that practicing the method on the basis of this description is prohibited. As will be demonstrated below, however, the method has promise as a treatment modality and, depending on one's practice, acquiring certification may well be worth it. Main steps/stages of the therapy are:

1. The method's essence is the exhortation and practice to be loud during production of progressively longer productions of "ah."
2. Another critical exercise is the increasingly competent and extended

- production of pitch change on “ah” or its equivalent, from the lowest to the highest possible pitch.
3. Furthermore, the emphasis is on helping the patient learn to identify the physical and perceptual cues associated with increasingly competent performance.
 4. The clinician’s tasks are to explain, encourage, systematically increase the production requirements, evaluate patient performance, give suggestions for altering performance as necessary, and systematically move the patient toward ever more competent performance.
 5. Counseling is critical because many patients question the mechanism of the treatment’s effect on swallowing.

Modifications

The above is a bare bones description of the methodology. The required training will introduce clinicians to the subtleties, and they are many. Only a sampling, plus a brief discussion of an intensive amplitude of movement treatment (Farley, Fox, Ramig, & McFarland, in press), appear below.

1. Voice quality must be monitored so as not to allow harsh, strained phonation.
2. The emphasis must be on increasing respiratory drive as normally as possible and not on squeezing the larynx unnaturally to prolong phonation.
3. Determining the duration and pitch change targets requires approaching the maximum the patient is capable of without deterioration in quality or consistent loudness.

4. Farley et al. (in press) describe the development of a treatment that emphasizes not only getting louder but also making bigger reaching and walking movements. They are calling this treatment “Big and Loud.” It, too, requires certification training. The essence of the method is that patients are given intensive practice in walking, reaching, and other daily tasks with greatly extended amplitudes. Early data are promising for speech and movement. Swallowing data are not apparently available.

Frequency and Duration

When used for speech, the prescription is for 16 sessions in 4 weeks with intensive homework. More, but seldom fewer, treatments may be necessary for dysphagic patients. Our dysphagia prescription is for a minimum of 16 weeks of home practice with one 1-hour clinic session scheduled every 1 or 2 weeks, depending on progress.

Candidacy

Those with PD would appear to be the best candidates; however, promising speech (swallow not evaluated) effects have also been reported for a limited number of patients with multisystem atrophy as well (Countryman & Ramig, 1994). We consider its use for all patterns of dysphagia in PD and in diagnoses where performance during swallowing seems generally underpowered or underdriven. Signs that may indicate this state (recognizing that other more focal conditions such as weakness may also produce similar signs) include slow bolus formation,

delayed initiation of the pharyngeal swallow with bolus trickling more or less continuously into the pharynx prior to initiation, reduced opening of the upper esophageal sphincter (UES), and residuals throughout the swallowing track. Depending on pattern, as when inadequate airway protection is a prominent sign, we may add other swallowing-specific techniques to be practiced in tandem. Those will be described below.

Effects

The data from a small group of PD patients to be reviewed below suggests a positive effect on behavioral plasticity. Specifically, rocking of the bolus, initiation of the swallow, and amount of postswallow residue were all reduced, and airway protection was enhanced. Admittedly, the mechanism or mechanisms remain speculative, but activation of wider areas of nervous activity in the early days followed by a progressive narrowing of the locus of nervous system activity (when the method is used to improve speech) is the pattern of neural activation (Liotti et al., 2003). The result may be the same for swallowing, but that remains an experimental question. Increased awareness may also be an explanation at a psychological level. Muscle strengthening seems unlikely but endurance may be increased. In the right clinician's hands, a placebo effect can also be predicted and, along with increased neural activation, may explain early therapeutic effects.

The Evidence

At the time of this publication, a single study on swallowing effects in move-

ment disorders, specifically PD, has been reported (El Sharkawi et al., 2002). As outlined above, LSVT is a promising treatment for dysphagia in PD that begs for further investigations.

RESPIRATORY MUSCLE STRENGTH TRAINING (RMST)

Description of Methods

RMST is of two main types: inspiratory (IMST) and expiratory (EMST). These two interacting respiratory muscle systems can be treated separately or simultaneously with resistance training. The training with the largest apparent effect requires the patient to forcefully inhale (IMST) or exhale (EMST) against graded, quantifiable resistance. A variety of commercial devices, such as that shown in Figure 14-1, are available for such training. Based on the amount of resistance provided, a certain amount of air pressure (measured in centimeters of water) is necessary to open the valve. If the resistance is systematically manipulated based on each patient's maximum strength, increased strength can be developed. Greater resistance and fewer repetitions are most effective for strength; less resistance and more repetitions may be more effective for endurance, although far less is known about appropriate prescriptions for endurance (Kays & Robbins, 2007). The methodology for strengthening as developed by Sapienza and colleagues (Kim & Sapienza, 2005; Saleem, Sapienza, & Okun, 2005; Sapienza, Davenport, & Martin, 2002; Silverman et al., 2006) is as follows.

15 Parkinson's Disease

This chapter, like all the disease chapters, is organized for easy review. It begins with definitions, followed by general signs and symptoms, epidemiology, evaluation, and treatment. Swallowing is featured in the second, longer section.

DEFINITION

Parkinson's disease (PD) is a chronic progressive neurodegenerative disease that begins in the brainstem and progresses to involve the entire brain (Braak, Ghebremedhin, Rub, Bratzke, & Del Tredici, 2004; Braak et al., 2003; Braak et al., 2006). However, the cardinal manifestations of bradykinesia/hypokinesia, rigidity, tremor, and postural instability are thought to be predominantly attributable to basal ganglia disease, including reduction of dopamine in the striatum, particularly the putamen (Bergman & Deuschl, 2002). PD is common, particularly in the elderly. In fact, among neurodegenerative conditions, only Alzheimer's disease is encountered more frequently (Nussbaum & Ellis, 2003). The cause of PD is unknown, but genetic and environmental influences (and their interactions) are thought to be involved. Most cases of PD occur sporadically, though a relatively small proportion of cases appear to be inherited.

SIGNS/DIAGNOSTIC CRITERIA

Main signs are: (a) bradykinesia/hypokinesia, (b) rigidity, (c) tremor, and (d) postural instability. *Bradykinesia/hypokinesia* refers to slowness in the onset and execution of volitional and nonvolitional movements, as well as a poverty of the amount and quality of movement. Behaviorally, these movement abnormalities may result in decreased arm swing with walking, *masked facies* (or *hypomimia*), and a shuffling, hesitant gait. Muscle *rigidity* occurs when the resistance of a joint to passive movement is increased. Patients will often describe a sensation of stiffness or an inability to relax their limb muscles. *Tremor* is often the most conspicuous sign of PD. Most typically, PD is associated with a resting tremor with a frequency of 4–7 Hz (Klockgether, 2004). Clinical criteria for diagnosis of PD usually include the presence of bradykinesia/hypokinesia with rigidity and/or tremor. In addition to the classic triad of PD signs is often added a fourth—*postural instability*. This sign is tested with the pull test, in which the clinician stands behind the patient and pulls back on his shoulders. Patients with postural instability will demonstrate a decreased ability to correct their posture and may

16 Multiple System Atrophy

DEFINITION

The definition and therefore diagnosis of multiple system atrophy (MSA) is at least modestly controversial, although a set of criteria for its confident or probable diagnosis has been published by Gilman and colleagues (1999). It is one of the typical forms of atypical parkinsonism. According to Gilman et al. (1999), MSA is a “progressive neurodegenerative disease of unknown etiology [which] occurs sporadically and causes parkinsonism with cerebellar, autonomic, urinary, and pyramidal dysfunction in many combinations” (p. 94). This heterogeneous picture led Shulman and colleagues (2004) to create the diagram that appears in Figure 16-1. In this diagram, the Parkinson features are labeled as extrapyramidal, the autonomic as autonomic failure, and the cerebellar as idiopathic cerebellar ataxia. This diagram ignores the corticospinal deficits but is useful nonetheless. It allows the capture of older nomenclature represented as OPCA for *olivopontocerebellar atrophy*, SND for *striatonigral degeneration*, and SDS for *Shy-Drager syndrome*. Gilman and colleagues (1999), reporting on the results of a consensus conference charged with specifying the criteria for a diagnosis of MSA, urged retirement of this terminology. Regarding Shy-Drager

syndrome, Gilman et al. stated that it is “no longer useful” (p. 198) as a label for the patient with severe autonomic dysfunction. Instead, Gilman and his group recommend the nomenclature MSA-P and MSA-C, depending on whether parkinsonian or cerebellar features predominate. Because the older terms SND for MSA-P and OPCA for MSA-C persist, and because at least a modest literature has appeared especially about OPCA, these terms will continue to appear along with MSA-P and MSA-C in this chapter.

SIGNS AND DIAGNOSTIC CRITERIA

For a diagnosis of MSA-P (or SND) to be entertained, evidence of one or more of the following is required: tremor, postural instability, rigidity, and bradykinesia or slowness of movement. In MSA-P, response to anti-PD medications is absent or short-lived. For MSA-C (or OPCA) to be diagnosed, ataxia of gait, gaze-evoked *nystagmus*, ataxic dysarthria, and ataxia of the lower limbs should be present on clinical examination. Autonomic dysfunction, which can predominate or occur in combination with other signs, includes impotence, orthostatic hypotension, and

18 Dystonia

DEFINITION

Dystonia is a heterogeneous condition that results in involuntary, sustained, and often repetitive contractions of opposing muscles resulting in twisting movements and abnormal postures. It is typical to classify dystonia by etiology (i.e., primary or secondary) or distribution (i.e., generalized, segmental, and focal). *Primary dystonia* may occur hereditarily or arise sporadically, while *secondary dystonia* is most commonly associated with other neurologic conditions (e.g., traumatic brain injury, anoxic brain injury). *Generalized dystonia* affects muscles throughout the body. In contrast, *focal dystonia* affects only one body part, such as the neck in cervical dystonia (or spasmodic torticollis) or the eyes in blepharospasm. *Segmental dystonia* affects two or more contiguous body parts, such as the eyes in combination with the lower face, mouth, and/or tongue in oromandibular dystonia (or Meige syndrome). Dystonia may also be classified based on age upon onset (early onset or late onset). Other classification systems and terminology may also be encountered in this complex condition, such as dystonia-plus syndromes (e.g., dopa-responsive dystonia, myoclonus-dystonia syndrome), pseudo-

dystonia, and psychogenic dystonia (Fahn, Bressman, & Marsden, 1998; Fernandez, Rodriguez, Skidmore, & Okun, 2007).

SIGNS/DIAGNOSTIC CRITERIA

The main signs are:

1. Twisting movements or abnormal postures of relatively long duration (in comparison to the brief movements encountered in chorea, for example).
2. Simultaneous contraction of opposing muscles in agonist and antagonist muscles in a given body part.
3. Though the involved muscle distribution may vary (and progress) over time, the affected musculature is fairly constant in comparison to the movement abnormalities encountered in patients with chorea.
4. Primary dystonia almost invariably begins with a single body part, often a leg or arm, and then gradually becomes more generalized.
5. Onset in the lower extremities tends to occur in younger individuals and be associated with a greater likelihood of progressing to generalized dystonia.