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Review Article

Single-Subject Investigation: A Model Oral-Motor Treatment Design for Children with Moebius Syndrome

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Abstract

This tutorial demonstrates the power and complexity of a single-subject treatment design for children with Moebius Syndrome (MS) who suffer from associated oral-neuromotor speech and swallowing difficulties. This suggested treatment protocol was constructed for a team of Speech-Language Pathologists (SLPs) interested in determining with confidence whether use of a unique oral stimulation tool actually results in reliable and valid improvements in speech intelligibility and swallowing in this target clinical population. This commercially available exercise tool was specifically designed to improve tongue weakness and incoordination in both children and adults with causally related neuropathologies or developmental oral-facial musculature immaturity. It has been used for several years by numerous SLPs with many different patient subgroups. However, virtually all highly supportive recommendations about its therapeutic utility have been anecdotal in nature. To date, no known comprehensive single-subject or group design scientific investigation has been conducted or published to corroborate these long-standing clinical impressions. The details of this single-subject design, along with the elaborate scoring and rating forms, can be creatively modified by investigators in other fields of study who wish to examine comprehensively the efficacy of any treatment technique for an intended patient or subject population.

Keywords: Moebius syndrome; Neuropathology; Velopharyngeal

Historical Perspectives of Case Study & Single-Subject Investigations

In a companion publication, alternative case study research designs and principles were reviewed in detail [1]. The current paper offers an example of a single-subject treatment protocol for a group of children with common clinical needs. Ostensibly, this research design resulted in a series of single-subject case study investigations. Whereas the conditions of treatment were directed at specific salient problems exhibited by the target population, the actual methods of data collection can be easily adapted and applied to any given study population. It is important to recall from the previous publication that a case series design is primarily an extension of the case study research format, wherein several subjects with conditions that closely resemble one another are examined in detail. These types of single-subject investigations are usually prospective in design, because they inherently require an inordinate amount of attention to detail and control during the data collection process. Not infrequently, results obtained from such study designs are based upon robust sets of data derived by close inspection of numerous tables and charts gathered throughout different phases of the treatment program. These supportive documents are deliberately designed by the researchers to illustrate the characteristic responses of a subject to specific treatments in the study protocol [2-6]. Notwithstanding such detail, more often than not, results of case series single-subject investigations do not usually lead to unequivocal conclusions, regardless of how strong the outcomes appear to be. Instead, because of design complexity and the limited number of subjects typically studied in any treatment program, these detailed investigations help develop supportive foundations for management of type-specific conditions. Most importantly, promising results often provoke theoretical considerations to replicate and expand upon the outcomes obtained through additional treatment efficacy research with other patients who have similar clinical needs.

As described previously, unlike a case study, a single-subject research design produces quantitative data from a single subject or group of single subjects; basically, each subject is treated as a solo experiment instead of one trial in a larger experiment. This method helps investigators comprehensively examine the possible causal relationship between specific interventions and behaviors of interest. However, generalizability to larger populations (i.e., external validity) remains limited. Minimizing extraneous or nuisance variables is of paramount importance to ensuring internal validity; that in a treatment paradigm nothing other than the independent variable of the study could reasonably explain observed improvements in the targeted dependent variable. In a single-subject investigation the sample size is usually limited to 1 to 10 participants, depending upon the study design. For this reason in particular, this small-N type of experimental research often meets the challenge of ensuring good internal validity. Subjects can serve as their own controls. On occasion, an observational group with similar diagnoses and clinical needs may be incorporated into this study protocol; that is, subjects on a waiting list who have not yet been enrolled in the treatment program. These individuals may be able to serve as external control subjects for comparative analyses with those undergoing treatment [7,8].

As discussed in the companion publication, any given single-subject investigation may take several months to gather sufficient

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data to complete the study. This lengthy process is in part due to the requirement to obtain a stable baseline profile prior to administration of an intended treatment. Such stability can be arbitrarily defined by the researcher at the outset, but usually includes at least several days of testing to be demonstrated. Data analyses in single-subject research designs differ from group research methodologies. In the latter, researchers typically employ various parametric and nonparametric inferential statistical computations to determine both the significance of experimental findings and whether the results can be generalized to a larger representation of the study population [9,10]. By contrast, single-subject research designs rely mostly on visual inspection of assorted data points represented on line graphs of one type or another. Use of certain statistical measures, such as percentages, means, and standard deviations can be employed in single-subject research to strengthen conclusions about treatment outcomes. Quantitative statistical computations such as a t-test or analysis of variance are not infrequently applied to analyze the data obtained and to determine the actual significance level of the treatment responses.

This paper describes an alternating treatment design with multiple probes across behaviors in a group of children with flaccid dysarthria and dysphagia secondary to Moebius Syndrome (MS). This single-subject experiment was originally constructed to investigate whether a unique sequence of tongue, jaw, and/or lip exercises prove therapeutically beneficial for this rare clinical population.

Background and Purposes of Study Proposal

First described by a German neurologist in 1888 MS is a rare, non-progressive, congenital disorder primarily characterized by lower motor neuron facial and eye movement paralyses [11]. Partial or complete paralyses can also involve the jaw, velopharyngeal, and tongue musculature in as many as 25% of cases. From patient to patient these pathophysiological features are highly variable in their presence and degree, and they are due to underdevelopment of those cranial nerves (V, VII, IX/X, XII) that normally innervate these oral-facial structures. Although the etiology remains elusive, the prevailing theory as to the possible cause of MS suggests that infantile nerve growth results from interrupted blood flow to the motor nuclei of the central nervous system along the brainstem during prenatal formation; genetic predisposition to this breakdown is also possible. A causal relationship between MS and the use of certain drugs during pregnancy, such as Misoprostol, Thalidomide, and Cocaine is similarly possible [12,13].

The incidence of MS is less than one per 500,000 births, with approximately 300 cases reported in the literature, and a prevalence of 2,000 cases worldwide. In addition to abnormal facial and eye appearances, owing to associated muscle weaknesses, children with MS frequently struggle with speech and swallowing difficulties secondary to variable disturbances of lip, tongue, jaw, and palatal movement patterns. In a smaller subset of children with MS coexisting limb deformities (e.g. clubfoot) and pectoral muscle hypoplasia may be observed. Most of the reported cases have been sporadic, and both sexes are affected equally.

The chief characteristics of MS include varying degrees of facial diplegia, sucking and swallowing abnormalities, articulation imprecision and unintelligibility, upper and lower limb fine and gross motor control disturbances, overall low muscle tone, cognitive deficits, and behavioral disorders. Research has focused primarily on the neurogenesis and treatment of MS, with no published prospective empirical investigations on neuro-rehabilitation strategies for prevailing oral-facial paresis and associated functional difficulties. Inasmuch as the majority of cases suffer from incomplete loss of muscle functions, the potential value of oral-neuromotor exercises for this population is of paramount interest and importance. The search for a reliable and effective treatment protocol for this unfortunate clinical population stems from well-documented reports that because children with MS often suffer from difficulties smiling, making facial expressions, swallowing, and producing intelligible speech they not infrequently experience emotional, psychological, and social stigmatization [14-16].

Exercises directed at the articulatory musculature, most notably those of the tongue, lips and mandible, have historically been applied for many different patient populations with the neurogenic motor speech disorder commonly referred to as dysarthria [17]. Whereas there are 7 different variants of dysarthria, depending upon the actual neuroanatomical site of abnormality, the pathophysiology of MS underscores the speech diagnosis of flaccid dysarthria in those individuals with articulation imprecision and slow-labored rate of speech. These abnormal features occur as a consequence of the aforementioned weaknesses of the facial, lingual, and/or jaw musculature. The underlying rationale for and characteristics of intensive motor speech exercises for patients with dysarthria has been studied for more than 50 years [17]. Experienced clinicians and researchers generally define oral-motor therapy as the process of facilitating the strength, coordination, range of motion, and tonal balance of all structures that compose the unified network of speech subsystems, including muscles of the oral mechanism, velopharyngeal and phonatory mechanisms, and lower respiratory tract.

There is a scarcity of empirical evidence that oral-neuromotor exercises actually help muscle functions in children with MS. As a consequence, there is a noticeable void in the rehabilitation literature regarding therapeutic methods that Speech-Language Pathologists (SLPs) should employ when working with this population to improve speech and swallowing abilities. Similar shortcomings exist in the speech rehabilitation literature for many other clinically needy individuals with dysarthria. To date the outcry for prospective, carefully designed, methodologically sound clinical research on the efficacy of oral motor exercises for children and adults with neuropathological speech and swallowing disorders, including those with MS, remains loud and unequivocal. This need for data-based investigations on the clinical relevance and value of an intensive physiological stimulation exercise program to improve speech and swallowing abilities in children with MS provoked the research proposal described below. Clinical researchers interested in conducting a similar study with a different patient population may find the detailed procedural descriptions helpful as they design their own IRB applications for approval.

Hypothesis of Current Investigation

The general null hypothesis of this investigation was as follows: For patients with MS and associated flaccid dysarthria and dysphagia sequential jaw-tongue physiology exercises, utilizing a specific stimulation therapy tool, do not result in significant improvement in articulation and/or feeding abilities. In support of this treatment focus is the well-known principle that early intervention can often provide individuals with disabilities the opportunity to improve daily living activities and quality of life by extinguishing developmental deficits and habitual dysfunction via proactive treatment strategies. Because of the rarity of MS, for the current investigation children of

all ages with clinically significant dysarthria and/or dysphagia will be recruited to participate so that a sufficient focused population can be studied. A primary outcome objective of this investigation is the establishment of a concentrated algorithm for the treatment of speech and swallowing difficulties, which can be reliably utilized by SLPs for their patients with MS.

Methods

Participants

Ten monolingual English-speaking children with the medical diagnosis of MS will be recruited to participate in this investigation. These individuals will be selected from a pool of prospective subjects. The diagnosis of MS will be based on the birth history of each prospective participant as well as confirmatory standard neurological medical examination results. Parental or guardian consent will be required and obtained in writing for each prospective participant.

Subject eligibility and exclusion criteria

To be included in this investigation, prospective participants will have to meet the following requirements:

1. Diagnosis of MS, as previous specified.
2. Dysarthria and dysphagia secondary to facial and lingual musculature weakness, as determined by preliminary baseline screening examinations to be performed by the anticipated treating clinician in this investigation.
3. Age five years or older.
4. No significant hearing loss.
5. No significant behavioral or cognitive impairments that may hamper treatment progress.
6. No history of concentrated oral motor exercises for dysarthria and/or dysphagia.
7. Parental or guardian consent to participate.
8. Mean rating below 80% on speech intelligibility baseline test results.
9. Rating of less than "6" on contextual speech baseline test results.
10. Mean rating of 3.0 or less on all dysphagia baseline test results.

Any one or more of the following factors will automatically exclude an individual from possible participation from this investigation:

1. Questionable diagnosis of MS.
2. Below five years of age.
3. Clinically insignificant speech or swallowing difficulties.
4. Hearing loss.
5. Autism.
6. History of oral-neuromotor speech or swallowing therapy.
7. Uncooperative parents or guardians.

Primary investigators

At least 2 experienced professionals will serve as co-principal investigators of this study. Each of these SLPs will have extensive clinical experiences treating children of all ages with MS and employing the oral motor therapy tools to be utilized in this investigation. These practitioners will recruit an expert team of SLPs who are specially trained in the application principles of the exercise protocols to be described below. The PIs will work collectively with this team of clinicians to evaluate each prospective subject to determine eligibility to participate in the study, with strict adherence to the aforementioned inclusion and exclusion criteria. It should be noted that the co-PIs and all SLPs who will be involved in the treatment of the participants of this investigation will have successfully completed all of the on-line Collaborative Institutional Training Initiative (CITI) courses.

Expert judges

A group of three blinded individuals, with expertise in the differential diagnosis and treatment of dysarthria and dysphagia, will be recruited and assembled to rate the participants (ie., MS subjects) on various anatomic and physiologic parameters pertaining to speech and swallowing functions. None of the judges will be familiar with the purpose of this investigation or the medical histories or identities of any of the subjects to be evaluated. The ratings to be rendered by the judges will be conducted independently according to the following sequence:

1. Prior to the start of the treatment program (i.e., baseline) for each subject.
2. Immediately after the last treatment session (i.e., @ discharge).
3. One month after completion of the entire therapy program (i.e., post-treatment).

Reliability measures

Inter-rater measures and analyses will be performed to ensure that judges are in agreement with each other on all diagnostic and treatment outcomes observations and conclusions 100% of the time. To control for intra-rater reliability the judges will be required to re-analyze a subset (i.e., 25%) of participants two weeks after completing their original pre- and post-treatment ratings. All such ratings by judges will be accomplished via independent viewing of digital video tapes. Specific scoring sheets and instructions for completion will be provided for each viewing task as described in detail below. Note: The treating clinicians will render all such ratings as well, for the purposes of subject selection and guidance during the therapy program. Their ratings, however, will not be calculated along with those of the independent expert judges for outcomes analyses.

Video tape procedures prior to treatment, at the completion of the treatment program, and after discharge from the program

All subjects will be videotaped using digital technology so that the expert judges can rate overall speech and swallowing difficulties, as described earlier. These tapes will be acquired (1) at baseline prior to initiation of the treatment program, (2) at discharge from the treatment program, and, (3) one month after the last treatment session.

Research design: baseline, maintenance, and generalization probes

To accept or reject the previously stated null hypothesis of this investigation, an alternating oral motor treatment design with multiple probes across behaviors will be employed. This design necessitates that all treatment parameters must initially undergo baseline testing, followed by non-continuous baseline measures to determine whether successfully treated behaviors are maintained. Conditions that have

yet to be addressed in the treatment protocol also continue to receive baseline testing. Training continues until all planned exercises have been directly administered and the subject has been discharged from treatment. A final probe session will be employed one month after the last treatment session to appraise carryover. The entire therapy program for each subject, in each group, will consist of 3 sessions per week for no more than six months from the start of the first therapy session.

Proactive Probes will be administered after treatment sessions 6, 12, 18, 24, 30, 36, 42, 48, 54, 60, 66, and 72 to track retention and possible transfer or generalization effects to untreated behaviors. These probes will be identical to the baseline measures using speech and swallowing scoring forms (Appendices). It should be noted that there may be subjects who make excellent improvements and meet the established criteria for discharge from therapy sooner than six months. In such cases, discharge will occur at that point in time. Conversely, those subjects who fail to meet early discharge criteria will remain in their respective treatment programs for the entire six months, as scheduled.

Variables of the investigation

The dependent variables, or treatment targets, will include two behaviors: (1) articulatory precision/intelligibility, and (2) oral feeding efficiency/safety.

The independent variables, or treatments themselves, will include an alternating oral motor exercise protocol of jaw-tongue physiology therapy using a specific stimulation tool.

Baseline measures

An initial baseline period with a minimum of three probe sessions (a), (b), and (c) over one week will be implemented for each subject prior to the initiation of any treatment protocol scheduled. Baseline stability will be required, as determined by rater judgments, before any subject will be permitted to proceed with his/her designated treatment protocol. Baseline data will be obtained on both dependent variables (i.e., treatment targets-speech intelligibility and swallowing efficiency).

Articulatory precision and overall speech intelligibility will be rated by judges as subjects perform three speaking tasks:

Task 1 will involve reading aloud ten separate index cards, each containing a prepared simple statement that ordinarily requires an answer by the listener. For example, "What number comes after seven? Subjects who are unable to read will be prompted by quiet and concealed verbal cues from the treating clinician to repeat the spoken statement.

Task 2 will involve speaking out loud the names of ten familiar picture cards (e.g., bus, car, rabbit, cookie, etc.) shown to them by the treating clinician.

Task 3 will involve describing out loud the events associated with an action picture.

For these three respective speaking tasks the judges will be requested to (1) answer each of the ten questions posed by the subject, (2) write the names of the ten pictures as spoken by the subject, and (3) rate overall articulatory precision and speech intelligibility of each subject during his/her action picture description.

Appendix A lists the ten questioning sentences and the names of the ten pictures to be spoken or named by the subjects. Appendix B illustrates the scoring form to be used by the judges to rate each subject's speech intelligibility performances. Note from this form that a 1-6 equal appearing interval scale will be adopted for all perceptual numerical ratings. At one extreme, a score of "1" will represent profound articulatory imprecision and unintelligibility. At the other end of the scale, a score of "6" will denote age appropriate (normal) speech characteristics. Similar ratings will also be rendered by the expert judges at the conclusion of the treatment program and at one-month post-treatment, as mentioned earlier.

Swallowing efficiency and safety will be rated by the judges as subjects perform three oral feeding tasks:

Task 1 will involve swallowing three teaspoons of 2% milk, administered sequentially by the treating clinician.

Task 2 will involve swallowing three teaspoons of apple sauce, administered by the treating clinician.

Task 3 will involve chewing and swallowing five cheerios followed by a chaser of 1 teaspoon of 2% milk, administered sequentially by the treating clinician.

For these respective oral feeding tasks judges will be requested to observe for several different aberrant oral motor and swallowing features, and to rate or record their presence or absence. After the completion of each of the three tasks the treating clinician will instruct the subject to voluntarily phonate the vowel /a/ for five seconds to enable the judges to rate the quality of voice and to discern perceptually the presence or absence of wet-gurgly voice quality; symbolic of possible liquid or food residue within the laryngeal inlet.

Appendix C represents the scoring form to be used by the judges to rate each subject's swallowing performances at baseline [18]. Note from this form the various tongue, lip, and jaw movement and postural features to be rated by the judges, as well as notable characteristics of bolus manipulation and ingestion difficulties, including drooling, frank or delayed coughing, choking, gagging, and gurgly voice behaviors after successive swallows. Similar ratings will also be rendered by the expert judges at the conclusion of the treatment program and one-month post-treatment.

The therapy program will only be initiated for subjects whose mean speech intelligibility baseline correct ratings on tasks 1 and 2 fall below 80%; task 3 mean scores across judges must be less than "6" (i.e., normal) in each case in order to begin the treatment program. Finally, to be eligible each subject's mean dysphagia baseline scores across judges must be 3.0 or less on each of the three tasks measured.

The treating clinicians will use their own perceptional ratings as comparative guidelines for determining the levels of improvement achieved throughout the therapy program, and for establishing whether to continue or discontinue specific exercises that are being employed in any given case. As mentioned earlier, bi-weekly probes (every six sessions) will be administered throughout the therapy program to facilitate comparisons with baseline levels of disability.

Therapy schedule & probe scoring forms during therapy

All participants will attend therapy three times a week for 30-60 minutes per session. Participants will be discharged from the intervention program according to their individual levels of progress achieved, or lack thereof, and at the discretion of the treating clinician in each case. A maximum of six months of therapy will take place for any given subject.

Therapy Program

Jaw-Tongue physiology therapy using the Lateralization-Elevation-Depression tool (LED)

Figure 1 illustrates the stimulation tool (LED) to be used in this treatment program to (a) stabilize and prohibit extraneous mandibular movements, and (b) facilitate improvements in 1) protrusion, 2) retraction, 3) depression, 4) elevation, 5) left lateralization, and 6) right lateralization movement control of the tongue [19]. These six target exercises will be introduced sequentially throughout the treatment program. Each will be treated every session, with equal time allotments per target per session. Their order of administration will be counterbalanced across the subjects in the group to reduce order effects. Appendix D illustrates the form that will be used by the treating clinicians each therapy session, throughout the treatment program, to rate the subject's performances during the tongue physiology tasks. Each exercise target may involve a different number of total trials per treatment session. The scoring format to be used by the treating clinician for each exercise target and each trial is as follows: The trial is to be rated as a composite performance. An "Acceptable" rating will be rendered for the trial only if the subject's physiologic performances throughout the entire trial were completely acceptable. If error movement patterns are observed for any of the attempts within a given trial that trial will be rated "Unacceptable".

At the completion of these trials each treatment session, the clinician will calculate the total number of "Acceptable" ratings rendered for each exercise trial/task and list the sum total next to the respective rating columns. The composite mean score of all trials per session will ultimately be transferred to a statistical log sheet as a single data point, which will represent the outcome of that treatment session.

Figure 1: Tongue lateralization and elevation therapy tool (TalkTools, Inc).

The LED technique

The hard rubber LED possesses grooves on each end that enable the user to bite down onto the block for jaw stabilization during various tongue physiology exercises. Placement of this tool varies within the mouth depending upon the target movement pattern. For tongue protrusion, retraction, and left/right lateralization exercises it is positioned between the subjects' upper and lower first molar teeth on either side of the mouth. Gentle biting pressure is encouraged for LED tasks. Visual biofeedback throughout all exercises will be achieved via a large desk mirror. For tongue elevation and depression exercises the LED is positioned between the upper and lower central incisor teeth. The criteria to be used by the treating clinician to rate all of these tongue movement performances as acceptable are as follows: (1) the target actions are performed to the fullest anatomical extent possible, and (2) all movements to and from the target positions are performed with smooth, graded, synchronous, rapid, and steady tongue control, without interruptive jaw activity.

For each of the aforementioned six movement tasks, numerous trials of 30 seconds each will be employed every treatment session; rest periods of 30 seconds will be permitted between trials. The order of the movement tasks will be altered every succeeding therapy session to reduce possible confounding order effects. The treating clinician will verbally provide all tongue movement prompts and instructions, with respect to when to start and when to stop for rest. As noted above, at the conclusion of each trial, the clinician will render a rating of the overall performance for that trial, using the form illustrated in Appendix D.

Exercise #1: Tongue protrusion: For this task, with the LED in place, the subject will be instructed to promptly and smoothly stick the tongue out as far as possible beyond the cutting edges of the upper and lower incisor teeth and then immediately pull it back into the mouth until the tip rests against the lower alveolar ridge. This physiologic round-trip will constitute one attempt and should be performed normally in no more than two seconds. Over the course of 30 seconds 15 such round-trip excursions of the tongue-tip should be possible, the total of which will be considered one trial. The resulting performance will be rated on the scoring form (Appendix D), as noted above. During each treatment session, 10 minutes of this protrusion exercise will be allotted, enabling up to 150 round-trips, including the intervening rest periods. In total, 10 trials will be possible per treatment session for this task.

Exercise #2: Tongue retraction: For this task, with the LED in place, the subject will be instructed to promptly protrude the tongue as far as possible and then forcefully, smoothly, and rapidly retract it into the mouth as far back as anatomically possible. This physiologic round-trip will constitute one attempt, and should be performed normally in no more than two seconds. Over the course of 30 seconds, 15 such round-trip excursions should be possible, the total of which will be considered one trial. The scoring form (Appendix D) will be used by the treating clinician to rate all such trials. During each treatment session 10 minutes will be allotted for this tongue retraction exercise, enabling up to 150 round-trips, including the intervening rest periods. A total of 10 trials will be possible per treatment session for this exercise.

Exercise #3: Tongue depression: For this task, with the LED in place, the subject will be instructed to use the tongue-tip to alternately press firmly against the small bead at the base of the tool, hold that position for five seconds, and then retract the tongue-tip off of the bead for five seconds. These alternate round-trip postures of the tongue-tip will be repeated six times, the total of which will be considered one trial, lasting one minute. A rest period of thirty seconds will be permitted between trials. During each treatment session 10 minutes will be allotted for this tongue-tip depression exercise, for a total of 7 trials. As many as 42 round-trips including rest periods will be possible per session, and the aforementioned scoring form (Appendix D) will be utilized by the treating clinician to rate and record the performances.

Exercise #4A: Mid-level tongue elevation: For this task, with the LED in place, the subject will be instructed to use the tongue-tip to press firmly against the bead at the base of the tool and raise it to the middle of the bar, to which it is attached, and then lower the bead to the base. This alternating tongue elevation/depression task will be performed five times in a row over the course of five seconds, or one

second per up/down round-trip. This performance will constitute one trial, after which a rest period of 25 seconds will be permitted. During each treatment session, 10 minutes will be allotted for this elevation/depression exercise, for a total of 20 trials. As many as 100 round-trip tongue-tip excursions, including rest periods, will be possible per session. The scoring form of Appendix D will again be used to rate each trial and to provide an overall mean score for the entire session, respectively.

Exercise 4B: Top-level tongue elevation: For this task, with the LED in place, the subject will be instructed to use the tongue-tip to press firmly against the bead at the base of the tool and raise it to the top of the bar to which it is attached and then lower the bead to the base. This alternating tongue elevation/depressions task will be performed five times in a row over the course of five seconds, or one second per up/down round-trip. This performance will constitute one trial, after which a rest period of 25 seconds will be permitted. During each treatment session, 10 minutes will be allotted for this elevation/depression exercise, for a total of 20 trials. As many as 100 round-trips, including rest periods, will be possible per session. The scoring form of Appendix D will again be used to rate each trial and to provide an overall mean score for the entire session, respectively.

Exercise 5A: Left tongue lateralization: For this task, with the LED between the upper and lower left first molars, the subject will be instructed to move the tongue to the left cheek area and use the tip to touch the bead at the bottom of the tool and hold that position for five seconds. After this performance, instructions will be given to return the tongue-tip to the central floor of the oral cavity and hold that position for five seconds. This task will be repeated six times in a row without a rest period. All six round-trip performances will take approximately one minute, and they will constitute one trial. After a succeeding rest period of 30 seconds, another six round-trip performances will be required, and so on until 10 minutes have elapsed for this exercise protocol. During each treatment session, a total of 7 trials will be possible, with as many as 42 round-trip left lateralization activities. The scoring form of Appendix D will be used to rate each trial and to provide an overall mean score for the entire session, respectively.

Exercise 5B: Mid-level left tongue lateralization: For this task, with the LED between the upper and lower left first molars, the subject will be instructed to move the tongue to the left cheek area and use the tip to touch the bead at the bottom of the LED and promptly raise it to the mid position on the bar, to which it is attached, and hold that position for five seconds. After this performance, instructions will be given to return the tongue-tip to the central floor of the oral cavity and hold that position for five seconds. This task will be repeated six times in a row without a rest period. All six round-trip performances will take approximately one minute, and they will constitute one trial. After a succeeding rest period of 30 seconds, another six round-trip performances will be required, and so on until 10 minutes have elapsed for this exercise protocol. During each treatment session a total of 7 trials will be possible, with as many as 42 round-trip left lateralization activities. The scoring form of Appendix D will be used to rate each trial and to provide an overall mean score for the entire session, respectively.

Exercise 5C: Top-level left tongue lateralization: For this task, with the LED between the upper and lower left first molars, the subject will be instructed to move the tongue to the left cheek area and use the tip to touch the bead at the bottom of the LED and promptly raise it to the top of the bar, to which it is attached, and hold that position for five seconds. After this performance, instructions will be given to return the tongue-tip to the central floor of the oral cavity and hold that position for five seconds. This task will be repeated six times in a row without a rest period. All six round-trip performances will take approximately one minute, and they will constitute one trial. After a succeeding rest period of 30 seconds, another six round-trip performances will be required, and so on until 10 minutes have elapsed for this exercise protocol. During each treatment session a total of 7 trials will be possible, with as many as 42 round-trip left lateralization activities. The scoring form of Appendix D will be used to rate each trial and to provide an overall mean score for the entire session, respectively.

Exercise 6A: Right tongue lateralization: For this task, with the LED between the right upper and lower first molars, the subject will be instructed to move the tongue to the right cheek area and use the tip to touch the bead at the bottom of the LED and hold that position for five seconds. After this performance, instructions will be given to return the tongue-tip to the central floor of the oral cavity and hold that position for five seconds. This task will be repeated six times in a row without a rest period. All six round-trip performances will take approximately one minute, and they will constitute one trial. After a succeeding rest period of 30 seconds, another six round-trip performances will be required, and so on until 10 minutes have elapsed for this exercise protocol. During each treatment session a total of 7 trials will be possible, with as many as 42 round-trip left lateralization activities. The scoring form of Appendix D will be used to rate each trial and to provide an overall mean score for the entire session, respectively.

Exercise 6B: Mid-level right tongue lateralization: For this task, with the LED between the upper and lower right first molars, the subject will be instructed to move the tongue to the right cheek area and use the tip to touch the bead at the bottom of the LED and promptly raise it to the mid position on the bar, to which it is attached, and hold that position for five seconds. After this performance, instructions will be given to return the tongue-tip to the central floor of the oral cavity and hold that position for five seconds. This task will be repeated six times in a row without a rest period. All six round-trip performances will take approximately one minute, and they will constitute one trial. After a succeeding rest period of 30 seconds, another six round-trip performances will be required, and so on until 10 minutes have elapsed for this exercise protocol. During each treatment session a total of 7 trials will be possible, with as many as 42 round-trip left lateralization activities. The scoring form of Appendix D will be used to rate each trial and to provide an overall mean score for the entire session, respectively.

Exercise 6C: Right tongue lateralization: For this task, with the LED between the upper and lower right first molars, the subject will be instructed to move the tongue to the right cheek area and use the tip to touch the bead at the bottom of the LED and promptly raise it to the top of the bar, to which it is attached, and hold that position for five seconds. After this performance, instructions will be given to return the tongue-tip to the central floor of the oral cavity and hold that position for five seconds. This task will be repeated six times in a row without a rest period. All six round-trip performances will take approximately one minute, and they will constitute one trial. After a succeeding rest period of 30 seconds, another six round-trip performances will be required, and so on until 10 minutes have

elapsed for this exercise protocol. During each treatment session a total of 7 trials will be possible, with as many as 42 physiologic round-trip left lateralization activities. The scoring form of Appendix D will again be used to rate each trial and to provide an overall mean score for the entire session, respectively.

Discontinuation of the jaw-tongue physiology therapy program

Discontinuation of the tongue exercise program will occur when all of the following criteria are met during any of the proactive baseline measurements (every sixth treatment session) throughout the length of the investigation: (1) the subject demonstrates at least 90% correct responses on speech intelligibility tasks 1 and 2, over three consecutive therapy sessions, (2) the subject demonstrates an improvement of at least two scale values from the initial baseline speech intelligibility rating on task 3, or a rating of "6" (normal) if the pre-treatment rating was "5", over three consecutive therapy sessions, and (3) the subject demonstrates an improvement of at least one scale value from the initial baseline swallowing proficiency ratings on all three tasks measured, or a rating of "4" (normal) if the pre-treatment rating was "3" on any of these tasks, over three consecutive therapy sessions. In the event the subject fails to achieve all three of the above criteria after respective probe data are obtained at treatment session #78 (26 weeks), the Jaw-Tongue Physiology treatment program will be discontinued and the subject will be discharged.

Discussion

The model single-subject treatment design described in detail in this paper illustrates the complex process of prospective data collection when conducting treatment research. Clinicians and patients alike will confront painstaking procedures when following the rules of this study design. Without strict adherence to these methods of data collection investigators will sacrifice both construct and internal validity of the data obtained throughout the investigation. As a consequence of faulty procedures, all conclusions drawn about the value of the findings obtained will be subject to deep skepticism.

When conducted properly single-subject treatment research methodologies, like the one proposed in this paper, offer investigators the opportunity to gather very reliable data relative to the efficacy of one or more therapeutic techniques; largely because these types of studies carefully define eligibility for participation as well as the criteria for sequential advancement from one exercise to the next within the protocol. Tight discontinuation regulations prohibit the treating clinician from continuously draining a subject to perform tasks that have proven too difficult to achieve. By tracking and rating subject performances throughout the protocol the treating clinicians and judges can delineate accurately whether to continue, alter, or stop any given treatment technique. When a subject responds well to a specific treatment the data tables and charts gathered by the investigators may not only confirm that improvements from baseline levels are notable, but these documents may also illustrate the exact nature and course of such gains over a specified period of time.

It has been the experience of this treating clinician that many children and adults with mild to moderate oral-neuromotor (i.e., tongue-lip-jaw) abnormalities are highly stimulable when exposed to rigorous, concentrated, and repetitive physiologic exercises aimed at improving fine and gross motor control of involved muscle groups [17-19].

Notable improvements often lead to additional gains as the treatment program advances to more complex tasks within the proposed exercise hierarchy. The data-based single-subject treatment design helps to produce reassuring evidence that the independent variables employed are responsible for the improvements observed in the dependent variables.

Unfortunately, those subjects with more severe muscle dyscontrol often struggle the most with specific exercise programs developed to improve function. Not infrequently, these individuals fail to reach criteria for advancement within the treatment program and are thus discharged relatively early in the course of management. Abiding by the methods of a structured single-subject treatment design the treating clinician is afforded the ability to identify and closely monitor these poor responders. The results obtained by such scrutiny enable cost-effective and efficient modifications in the program for these individuals, with hopes that adjustments may prove more beneficial than the originally administered treatments.

Conclusion

Single-subject studies often produce robust data-bases. The prospective rigor of obtaining these data vary in complexity depending upon the type of single-subject research design employed. In this paper, an alternating treatment design with multiple therapeutic probes was proposed as a model for clinical researchers who might be contemplating a single-subject investigation to determine the efficacy of a specific treatment for a particular clinical population. The target study group of this model investigation was children with MS and commonly associated flaccid dysarthria and dysphagia. The data collection techniques and rating forms elaborately described and illustrated how to collect and record subject performances throughout the experiment to ensure validity of the results reported.

Unfortunately, the clinical scientific literature is not replete with investigations that have adhered to such rigorous standards for data collection, reliability, and validity; probably because doing so is extremely time consuming and financially impractical for most clinicians. The scientific literature in all fields of study cannot produce clinically useful information if contributing researchers conduct studies without sound scientific methods of data collection. As this paper attempts to demonstrate, even the results of a single-subject investigation can prove quite convincing and clinically valuable when the data to be gathered are collected systematically and with respect for the inherent demands and standards of an accepted research design.

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Appendix A

Speaking Task #1 (Questioning sentences)

1. What number comes after seven?
2. Who was the first president of the United States?
3. What do you like to drink with chocolate-chip cookies?
4. What animal likes to bark?
5. When birds fly, what do they flap to lift off the ground?
6. In what month do people celebrate Christmas?
7. Which season is the coldest?
8. How many nickels are in a quarter?
9. What do we use to clean our teeth?
10. What does a man use to shave his face?

Speaking Task #2 (Picture cards)

1. Computer
2. Puppy
3. Shovel
4. Cheese
5. Lollipop
6. Zebra
7. Nose
8. Sun
9. Rabbit
10. Telephone

Speaking Task #3 (Picture description)

Any Action Photograph May Be Used Here

Appendix B

Articulation Precision/Speech Intelligibility Scoring Form

Instruction #1: The first set of speech samples represent 10 different questioning statements by the speaker. As judge you are required to answer each of the 10 questions in the respective spaces below:

Answer #1	_____	Answer #6	_____
Answer #2	_____	Answer #7	_____
Answer #3	_____	Answer #8	_____
Answer #4	_____	Answer #9	_____
Answer #5	_____	Answer #10	_____

(# correct X 10 = ____%)

_____

Instruction #2: The second set up speech samples represent 10 different picture naming performances by the speaker. As judge, you are required to list the names of each of the 10 pictures named by the speaker in the respective spaces below:

Word #1	_____	Word #6	_____
Word #2	_____	Word #7	_____
Word #3	_____	Word #8	_____
Word #4	_____	Word #9	_____
Word #5	_____	Word #10	_____

(# correct X 10 = ____%)

_____

Instruction #3: The third speech sample represents the speaker's description of a picture scene. As judge, you are required to use the 6-point equal appearing interval scale below to rate your perceptions of the speaker's overall articulation precision and speech intelligibility. Any voice difficulties should not be considered in your rating. Note that at the extremes of this scale a rating of "1" would represent profound speech difficulty, and a rating of "6" would indicate generally normal or unremarkable speech precision and intelligibility. Circle the numeral that reflects your perceptions of the speaker.

1	2	3	4	5	6
Profound speech difficulty	General speech difficulty	Moderate speech difficulty	Mild to moderate speech difficulty	Mild Speech difficulty	Normal speech

Appendix C

Dysphagia Scoring Form

Instructions: Use this rating scale to evaluate the presence (or absence) of specific swallowing difficulties and their level of severity. Rate all three swallow tasks and all associated feature categories by circling their respective numeric ratings. Do not render a composite rating until all swallow attempts are completed for each of the three tasks and the subject has been requested to prolong the vowel /a/ for 5 seconds.

Task #1 (3X5cc Milk swallows)

A. Signs of Coughing

1	2	3	4
Immediate, persistent and severe	Delayed persistent and moderate	Subtle and inconsistent	None

B. Signs of Drooling

1	2	3	4
Severe, persistent and bilateral	Moderate, persistent and unilateral	Subtle and inconsistent	None

C. Signs of Nasal Regurgitation

1	2	3	4
Persistent and Severe	Moderate and persistent	Subtle and inconsistent	None

D. Signs of Throat Clearing

1	2	3	4
Profound and persistent severe	Moderate and persistent	Subtle and inconsistent	None

E. Sign of Tongue Thrusting

1	2	3	4
Severely evident	Moderately evident	Mildly evident	None

F. Signs of Lip Closure Incompetency

1	2	3	4
Severely evident	Moderately evident	Mildly evident	None

G. Signs of Chewing or Mandibular Incoordination

1	2	3	4
Severely evident	Moderately evident	Mildly evident	None

H. Signs of Wet-Gurgly Voice Quality

1	2	3	4
Severely evident	Moderately evident	Mildly evident	None

Task #2 (3X5cc Applesauce Swallows)

A. Signs of Coughing

1	2	3	4
Immediate, persistent and severe	Delayed persistent and moderate	Subtle and inconsistent	None

B. Signs of Drooling

1	2	3	4
Severe, persistent and bilateral	Moderate, persistent and unilateral	Subtle and inconsistent	None

C. Signs of Nasal Regurgitation

1	2	3	4
Persistent and Severe	Moderate and persistent	Subtle and inconsistent	None

Appendix C (Cont'd)

D. Signs of Throat Clearing

1	2	3	4
Profound and persistent severe	Moderate and persistent	Subtle and inconsistent	None

E. Signs of Tongue Thrusting

1	2	3	4
Severely evident	Moderately evident	Mildly evident	None

F. Signs of Lip Closure Incompetency

1	2	3	4
Severely evident	Moderately evident	Mildly evident	None

G. Signs of Chewing or Mandibular Incoordination

1	2	3	4
Severely evident	Moderately evident	Mildly evident	None

H. Signs of Wet-Gurgly Voice Quality

1	2	3	4
Severely evident	Moderately evident	Mildly evident	None

Task #3 (5 cheerios + 5cc 2% milk)

A. Signs of Coughing

1	2	3	4
Immediate, persistent and severe	Delayed persistent and moderate	Subtle and inconsistent	None

B. Signs of Drooling

1	2	3	4
Severe, persistent and bilateral	Moderate, persistent and unilateral	Subtle and inconsistent	None

C. Signs of Nasal Regurgitation

1	2	3	4
Persistent and Severe	Moderate and persistent	Subtle and inconsistent	None

D. Signs of Throat Clearing

1	2	3	4
Profound and persistent severe	Moderate and persistent	Subtle and inconsistent	None

E. Signs of Tongue Thrusting

1	2	3	4
Severely evident	Moderately evident	Mildly evident	None

F. Signs of Lip Closure Incompetency

1	2	3	4
Severely evident	Moderately evident	Mildly evident	None

G. Signs of Chewing or Mandibular Incoordination

1	2	3	4
Severely evident	Moderately evident	Mildly evident	None

H. Signs of Wet-Gurgly Voice Quality

1	2	3	4
Severely evident	Moderately evident	Mildly evident	None

Adapted from McHorney et al. 2002 **Page 2 of 2**

Appendix D

Composite Rating Form for Tongue Physiology Exercises

Instructions: Use this form to rate tongue exercise performances per trial as follows: An "A" rating represents acceptable performances, and a "U" rating denotes an unacceptable effort. Acceptable indicates that the tongue postural adjustment being rated was smooth, complete, synchronous, rapid, and without interruptive hyperactive jaw activity. Unacceptable indicates the presence of any one or more of the following slow-labored, incomplete range of motion, dysrhythmic and interruptive tongue or jaw movement patterns. When calculating the overall percentage of acceptable performances, all "A" ratings will receive a score of "1" and all "U" ratings will receive a score of "O".

Place either an "A" or "U" in the box under the trial # being scored.

Exercise #1: Protrusion (15 trials X 10 repetitions per trial)

1 2 3 4 5 6 7 8 9 10 11 12 13 14 15

Exercise #2: Retraction (15 trials X 10 repetitions per trial)

1 2 3 4 5 6 7 8 9 10 11 12 13 14 15

Exercise #3 Depression: (7 trials X 6 repetitions per trial)

1 2 3 4 5 6 7

Exercise #4a: Mid-Level Elevation (20 trials X 5 repetitions per trial)

1 2 3 4 5 6 7 8 9 10

11 12 13 14 15 16 17 18 19 20

Exercise #4b: Top-Level Elevation (20 trials X 5 repetitions per trial)

1 2 3 4 5 6 7 8 9 10

11__ 12__ 13__ 14__ 15__ 16__ 17__ 18__ 19__ 20__

Appendix D (Cont'd)

Exercise #5a: Left-Lateral (7 trials X 6 repetitions per trial)

1 2 3 4 5 6 7

Exercise #5b: Left Mid-Level Lateral (7 trials X 6 repetitions per trial)

1 2 3 4 5 6 7

Exercise #5c: Left Top-Level Lateralization (7 trials X 6 repetitions per trial)

1 2 3 4 5 6 7

Exercise #6a: Right Lateral (7 trials X 6 repetitions per trial)

1 2 3 4 5 6 7

Exercise #6b: Right Mid-Level Lateralization (7 trials X 6 repetitions per trial)

1 2 3 4 5 6 7

Exercise #6c: Right Top-Level Lateralization (7 trials X 6 repetitions per trial)

1 2 3 4 5 6 7