Pediatric Neurology 000 (2018) 1–7

Contents lists available at ScienceDirect

Pediatric Neurology

journal homepage: www.elsevier.com/locate/pnu

Research Paper

# Home-Based, Therapist-Assisted, Therapy for Young Children With Primary Complex Motor Stereotypies

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ARTICLE INFO

*Article history:*
Received 26 March 2018
Accepted 6 May 2018

*Keywords:*
movement disorder
motor stereotypies
behavioral therapy
home-based therapy
Stereotypy Severity Scale

ABSTRACT

*BACKGROUND:* Complex motor stereotypies (CMS) typically begin before age three years and include rhythmic, repetitive, fixed movements that last for seconds to minutes and can be interrupted with distraction.

*OBJECTIVE:* We evaluated the effectiveness of a home-based, parent-provided therapy accompanied by scheduled telephone calls with a therapist, in five- to seven-year old children with primary CMS.

*METHODS:* Eligible families received an instructional digital versatile disk (DVD) written instructions, and scheduled telephone contacts with a therapist at baseline (DVD receipt), one, three, and eight weeks later. At each call, parents completed outcome measures and received feedback. Outcome scales Stereotypy Severity Scale (SSS) Motor and Impairment scales and a Stereotypy Linear Analogue Scale (SLAS) were also completed via the Iinternet (REDCap)—at screening, one and two months post-baseline call. At study conclusion, participants were divided into an intent-to-treat (ITT; had at least one call) or a lost-to-follow-up (LTF) group.

*RESULTS:* Thirty-eight children (mean = 5 years ± 11 months) were enrolled. The LTF group (n = 14) had significantly higher scores than the ITT (n = 24) group on all attention-deficit/hyperactivity disorder ratings ($P < 0.01$), but not stereotypy severity. Primary outcome scores, acquired by telephone and REDCap, showed a significant reduction in SSS Motor and Impairment scores between the initial and the last completed evaluation ($P \leq 0.001$). Calculated change ratios were SSS Motor −0.23/−0.30 (cal/REDCap); SSS Impairment −0.31/−0.32; and SLAS −0.54 (REDCap). Clinical improvement was further supported by results from a parent improvement scale and end of study questionnaires.

*CONCLUSIONS:* Home-based, parent-administered behavioral therapy supplemented by telephone contact with a therapist is effective in reducing complex motor stereotypies in children.



## Introduction

Motor stereotypies are repetitive, rhythmic, fixed, purposeful but purposeless movements that stop with distraction.[1] Complex motor stereotypies (CMS) typically involve the upper extremities with hand and/or arm flapping or waving, wrist flexion and extension, hand opening and closing, and finger wiggling. Additional accompanying movements often include jumping, pacing, mouth opening, head posturing, and occasionally vocalizations. Movements typically begin in early childhood

Conflict of interest: The DVD discussed in this research is available for purchase via the Johns Hopkins Health Care Solutions (https://www.johnshopkins-solutions.com/solution/the-johns-hopkins-motor-stereotypies-behavioral-therapy-program). Proceeds go to the Johns Hopkins Motor Stereotypies Research Fund to support further investigations on this disorder.

Financial support: This research was supported by U54HD079123.

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https://doi.org/10.1016/j.pediatrneurol.2018.05.004
0887-8994/

and tend to occur when excited, engrossed, fatigued, or stressed. They occur multiple times per day, and last for seconds to minutes, or even longer.[2] CMS are divided into two groups: "primary" for those who are otherwise developmentally normal and a "secondary" category for those with developmental issues, e.g. autism, Rett syndrome, and inborn errors of metabolism. A follow-up study in teens and young adults with primary CMS has shown that the frequency and duration of movements diminish, but usually persist.[3] Pathophysiologically, stereotypies are hypothesized to arise from alterations within habitual motor pathways within the brain.[4]

Children with primary motor stereotypies frequently deny concerns or physical issues; however, there are ongoing worries about social stigmatization, classroom disruption, or the possibility of interference with academic activities. Pharmacological therapy, based on parent report of prior medication trials, has not identified an effective agent.[5] In contrast, behavioral therapy has been shown to be beneficial.[6,7] In 12 children, ages 6 to 14 years with primary CMS, therapist-based training using a combination of awareness and differential reinforcement of other behaviors was successful in reducing movements.[6] A second study evaluated the efficacy of an instructional digital versatile disk (DVD) as a home-based, parent-administered behavioral therapy[7]—the latter contained instructional approaches successfully utilized in the therapist protocol. At their final post-DVD assessment, 54 children, aged 7 to 14 years, showed a significant improvement, as compared with baseline, on all three primary assessment measures. One noteworthy limitation, recognizing that stereotypies typically begin before the age of three years, was the focus of therapy on older (7-14-year old) children. A further problem with the later trial was the large number of enrolled dropouts (64%); both before and following, the initial parent completed evaluation.

The goal of the current study was twofold: (1) to evaluate behavioral therapy in children with primary CMS ages five to seven years; and (2) to determine the benefit of a home-based, parent-administered behavioral therapy administered in conjunction with telephone support provided by a knowledgeable behavioral therapist. We hypothesized that this combined home-based DVD-therapist support approach would enhance training, diminish the number of dropouts, and reduce stereotypies in a younger population of children with CMS.

## Methods

### *Overview*

This was a Johns Hopkins Medicine Institutional Review Board approved protocol. Children with primary CMS were recruited from either the Johns Hopkins Pediatric Neurology Movement Disorder Outpatient Clinic (HSS, Director), or via e-mail (singerlab@jhmi.edu). All participants verbally consented and each participant's parents consented in writing. Study flow is outlined in the Figure. In brief, the study coordinator (SR), using standardized forms completed a brief general history, obtained baseline data about each child's stereotypies, and completed an Autism Spectrum Screening Questionnaire (ASSQ). The presence of stereotypic movements was confirmed, either via direct observation in clinic or by video review (HSS). Additional baseline assessments included primary outcome measures (Stereotypy Severity Scale—SSS, Motor and Impairment scores; Stereotypy Linear Analog Scale—SLAS), a secondary outcome measure (Patient Global Impression of Improvement—PGI-I), and comorbidity measures (Multidimensional Anxiety Scale for Children—MASC; attention-deficit/hyperactivity disorder (ADHD)-Rating Scale IV; Conner's Parent Rating Scale—CPRS; Repetitive Behavior Scale-Revised—RBS-R; Children's Yale-Brown Obsessive Compulsive Scale—CYBOCS; and Social Responsiveness Scale—SRS). All of the aforementioned were completed by parents on REDCap; an electronic web-based application for data capture and online questionnaires. Once screening and baseline scales were completed, the training DVD, written instructions, and a log sheet for tracking the behavioral therapy were mailed. After confirmation of receipt of the DVD, the behavioral therapist (HRW) contacted the family via telephone (baseline call). Subsequent therapist contacts occurred at one, three, and eight weeks following the baseline call. At each call, in addition to educational discussions (described below), stereotypy rating scores were obtained. Independently completed parent rating scales, via REDCap, were obtained before receipt of the DVD, at four and eight weeks after the baseline call. Upon completion of the protocol, the parents were asked to finalize an end of study form.

### *Subjects*

A total of 38 children (24 boys, 14 girls), ages five to seven years (mean 5 years ± 11 months) with primary CMS successfully completed screening and received a DVD. Eligibility required participants to have: (1) confirmed CMS; (2) onset before the age of three years; (3) no reported premonitory urge; and (4) temporary suspension of movements by an external stimulus or distraction. Exclusion criteria included: (1) an abnormal total score (greater than 13) on the ASSQ or a prior autism spectrum disorder diagnosis; (2) evidence of intellectual disability; (3) seizures or a known neurological disorder; and (4) the presence of motor/vocal tics. The presence of inattentiveness, hyperactivity, or impulsivity (i.e., ADHD symptoms) and/or obsessive compulsive behaviors were not exclusionary. Participants on medications were allowed to continue on prescribed medications at a stable dose for the duration of the study. A total of 15 children had been evaluated in the Pediatric Neurology Clinic at Johns Hopkins for repetitive movements before participation in this study.

### *Behavioral therapy*

(1) Parent-directed video-based behavioral therapy

The instructional DVD (44 minutes) included a 10-minute didactic on CMS followed by instructions provided by a behavioral psychologist and instructional vignettes.[7] Parents were instructed to implement awareness training in the first week with the addition of data collection and reinforced suppression in week two and beyond.

(a) *Awareness training*: The goal of awareness training is to make the child aware of his/her movements through the use of videos showing the activity and the practice of voluntarily starting and stopping the movement. During these sessions, parents provided positive verbal reinforcement (e.g., good job, that is right) when the behavior closely approximated the actual behavior. If the child's behavior did not closely approximate the behavior, parents were instructed to correct the child by either mimicking the behavior themselves and/or verbally asking the child to modify the behavior (e.g., flap your hands faster, open your mouth). Practice sessions were twice daily, and included five repetitions of doing the movement for 30 seconds followed by a one-minute rest period. If the child resisted practicing, parents were asked to try to make some preferred activity (e.g., TV, video game access) contingent on completing the trial.

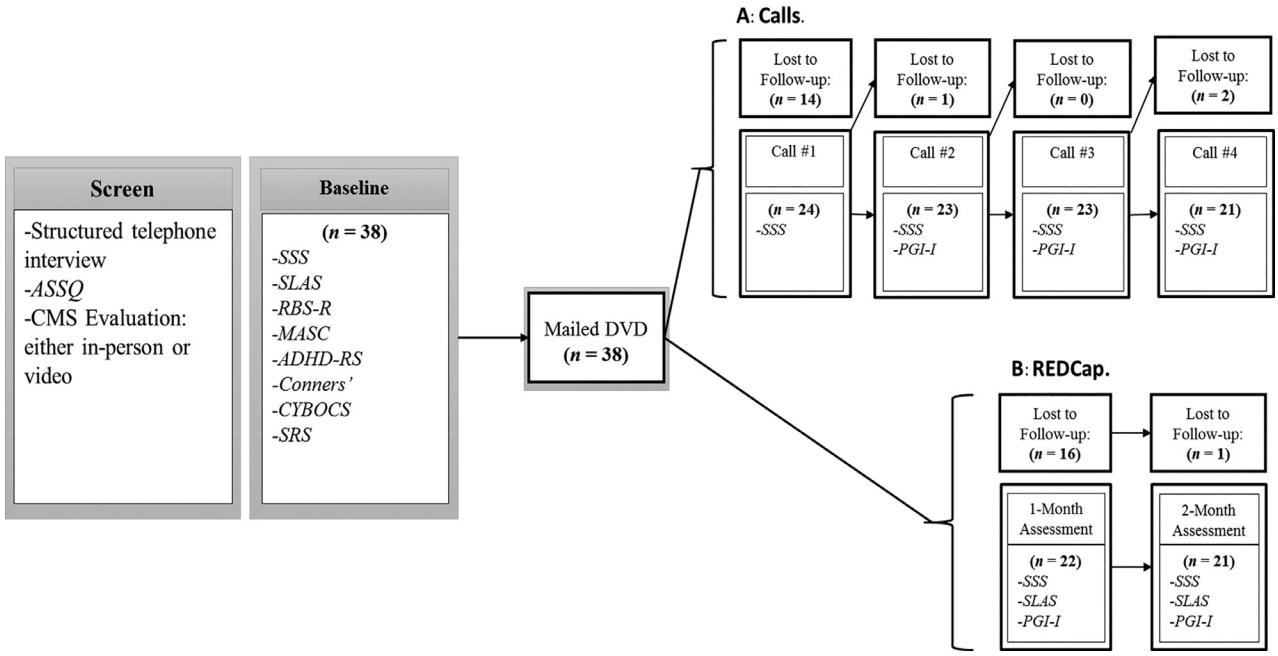


**FIGURE.**
Study flow chart for calls and REDCap ratings.

(b) *Differential reinforcement of other behavior (DRO)* or *reinforced CMS suppression*: The goal of DRO was the suppression of motor stereotypies. Requested practice sessions occurred five to ten times per day, with initial session durations of one to five minutes gradually increasing to 30 minutes. Practice sessions were conducted in different situations, focusing on those where the behavior is more likely to occur. Parents were instructed to deliver frequent verbal reinforcement during the interval when the behavior was not occurring and to provide reinforcements (prizes, rewards).

(2) Telephone calls with the behavioral psychologist

Telephone calls were made to parents on four occasions: (1) baseline—after receiving and indicating that they had watched the DVD; (2) one week later; (3) two weeks after the second call; and (4) five weeks after the third call. Calls typically lasted 15 to 30 minutes. Most calls were conducted with only one parent. Each call began with administering the SSS and the PGI-I.

Call #1 (baseline call): The overall goals of the training program were reviewed with an initial emphasis on the importance of awareness training. The duration, location, and the significance of practice sessions were discussed and suggestions provided regarding its implementation. Parents were requested to maintain daily behavior logs to help determine situations in which the behavior was likely to occur and to ignore comments about stereotypies present outside of practice sessions.

Call #2 (one week later): Data regarding awareness training were reviewed and schedules adjusted. DRO was reviewed and practice sessions discussed; initially using short intervals so that at least a 95% success rate could be achieved. The necessity for frequent verbal reinforcement and use of tangible reinforces were emphasized.

Call #3 (study week three): Data were collected on practice frequency of awareness training (discontinued at Call #3) and DRO—adjustments were made to assist in attaining a 95% successful period of behavioral suppression during each DRO session. For children in school or a structured program, the possibility of at least one DRO session per day at that location was recommended. If performed, parents were requested to obtain feedback from the administrative staff and provide reinforcement.

Call #4 (study week eight): Data were collected on the frequency of DRO practice and further adjustments were suggested about the ongoing use of the training program.

### *Assessment measures*

The SSS is a five-item caregiver questionnaire consisting of two components (Motor and Impairment) for the ranking of motor stereotypy severity.[6,8] The SSS Motor score (range 0-18) quantifies motor severity and rates movements along four discriminate dimensions: number (0-3), frequency (0-5), intensity (0-5), and interference (0-5). The SSS Impairment score (range 0-50) is an independent rating of difficulties in self-esteem, family, school, or social acceptance caused by the movements.

The SLAS is a 100-millimeter continuous line on which the parent places a mark indicating their child's stereotypy activity, ranging from 0 (best it has ever been) to 100 (worst it has ever been), considering the dimensions number, frequency, intensity, and interference.

The PGI-I is a seven-item scale that asks the parent to rate the relative improvement of CMS experienced by the patient since the beginning of the study. Ratings include: 1 (very much better), 2 (much better), 3 (improved), 4 (no change), 5 (minimally worse), 6 (much worse), and 7 (very much worse).[6]

The ASSQ is a 27-item caregiver questionnaire addressing symptoms of autism spectrum disorders. Children with total ASSQ scores greater than 13 were excluded.[9]

Other parent completed measures included: the MASC, assessing symptoms of anxiety[10]; the ADHD-Rating Scale-IV[11] and CPRS,[12] assessing symptoms of ADHD; the CYBOCS, assessing symptoms of obsessive compulsive disorder (OCD)[13]; the RBS-R, assessing repetitive behaviors,[14] and the SRS, assessing social communication skills.[15]

### *Statistical analyses*

After screening, a total of 38 participants completed the baseline assessment and received the training DVD. This aggregate

population was then subdivided into (1) the *lost-to-follow-up (LTF) group* and (2) the *intent-to-treat (ITT) group.*

(1) The LTF group included those participants who completed no post-DVD receipt assessments. This group contained 14 participants, nine boys and five girls. This subsample was 86% Caucasian, 7% Hispanic, and 7% Asian. One participant reported the use of dexmethylphenidate (Focalin XR) and methylphenidate for ADHD. Parents of eight participants in the LTF group withdrew their child from the study after receiving the DVD, citing one of the following reasons: mild symptoms, associated time commitment, extenuating family circumstances, or child anxiety about the treatment. Five parents gave no reason. The study coordinator withdrew one participant due to failure to respond to multiple weekly telephone prompts.
(2) The ITT group included participants who completed at least one phone call with the study psychologist, after receiving the DVD. This group contained 24 participants, 15 boys and 9 girls. This subsample was 84% Caucasian, 4% Hispanic, 4% American Indian, and 8% Asian. No participants in the ITT group were receiving psychotropic medications or reported concurrent or prior pharmacological or behavioral treatment for CMS.

Comparisons were made between participants in the LTF and ITT groups for demographic variables (age, age of stereotypy onset, sex, race and/or ethnicity, prior treatment, current medication status, stereotypy severity ratings, and scores on comorbidity measures) using *t* tests for continuous variables, and chi-square analyses for categorical variables. For group comparisons, assumptions for parametric analyses were assessed using Kolmogorov-Smirnov tests for normality of distributions, with non-parametric analyses used as indicated. Within the ITT group, repeated ANOVA measures between the baseline and last assessment were performed to evaluate the level of change. Finally, the outcome assessments from the last rating made by telephone call were directly compared with the last ratings made by parents in REDCap (n = 22) to assess comparability of the two assessment formats.

## Results

### *Group (LTF and ITT) comparisons at baseline*

Baseline characteristics were compared between the LTF and ITT groups in order to determine whether there were meaningful differences. Results for continuous variables are listed in Table 1. The LTF group had significantly higher scores on all ADHD ratings (CPRS and ADHD Rating Scale-IV), compared with the ITT group, indicating that those with greater ADHD symptomatology at baseline were more likely to drop out. For the ADHD Rating Scale-IV, mean scores for both Hyperactivity/Impulsivity and Inattention in the ITT group were in the average range for age, while mean scores for both scales in the LTF group were mildly elevated for age. Otherwise, there were no significant group differences in terms of baseline age, age of stereotypy onset, sex [$\chi^2_{(2)} = 0.31$, $P = 0.728$], race [$\chi^2_{(4)} = 0.75$, $P = 0.860$], maternal education [$\chi^2_{(6)} = 5.77$, $P = 0.329$], ASSQ scores, and parent-completed screening REDCap SSS Motor or Impairment scores, SLAS ratings, CYBOCS, MASC, RBS-R, or SRS scores. Of note, mean scores for Total SRS for both ITT and LTF groups were within normal limits for age.

**TABLE 1.**
Baseline Differences Between Intent-to-Treat and Lost-to-Follow-Up Groups

| Measure | ITT | | LTF | | | |
|---|---|---|---|---|---|---|
| | n = 24 | | n = 14 | | | |
| | Mean | SD | Mean | SD | *P* | $\eta^2$ |
| Age at study (years) | 5.96 | 0.81 | 5.93 | 0.83 | 0.914 | 0.000 |
| Age of onset (months) | 12.17 | 10.16 | 11.36 | 11.73 | 0.824 | 0.001 |
| ASSQ total | 6.46 | 3.40 | 6.71 | 3.27 | 0.822 | 0.001 |
| CPRS Inattention | 6.58 | 4.75 | 11.57 | 8.23 | ***0.023*** | 0.136 |
| CPRS H-I | 11.63 | 7.09 | 18.21 | 11.30 | ***0.033*** | 0.120 |
| ADHD RS Inattention | 5.83 | 4.54 | 11.36 | 9.16 | ***0.017*** | 0.147 |
| ADHD RS H-I | 7.17 | 5.07 | 12.36 | 8.66 | ***0.025*** | 0.132 |
| CYBOCS obs | 0.92 | 1.64 | 0.38 | 1.39 | 0.328 | 0.027 |
| CYBOCS com | 0.83 | 2.28 | 1.15 | 2.88 | 0.712 | 0.004 |
| MASC total | 66.58 | 13.54 | 73.00 | 14.72 | 0.181 | 0.049 |
| RBS-R total | 8.00 | 5.21 | 9.36 | 8.54 | 0.545 | 0.010 |
| SRS total | 28.63 | 14.46 | 35.93 | 22.42 | 0.256 | 0.036 |
| SLAS total | 52.79 | 26.51 | 51.71 | 30.07 | 0.909 | 0.000 |
| SSS Motor | 12.08 | 1.72 | 11.64 | 2.06 | 0.483 | 0.014 |
| SSS Impairment | 16.25 | 8.75 | 17.14 | 9.14 | 0.767 | 0.002 |

Abbreviations: ADHD-RS =ADHD Rating Scale IV, Home Version; ASSQ = Autism Spectrum Screening Questionnaire; Com = compulsive; CPRS = Conners' Parent Rating Scale, Third Edition; CYBOCS = Children's Yale-Brown Obsessive Compulsive Scale; H-I = hyperactivity/impulsivity; ITT = Intent-to-Treat; LFT = Lost-to-Follow-up; MASC = Multidimensional Anxiety Scale for Children; Obs = obsessive; RBS-R = Repetitive Behavior Scale-Revised; SLAS = Stereotypy Linear Analog Scale; SRS = Social Responsiveness Scale; SSS = Stereotypy Severity Scale.

### *Intent-to-treat group analyses*

Within the ITT group, neither age at study entry nor age of stereotypy onset was significantly associated with any of the baseline stereotypy ratings (all $P > 0.23$). At baseline, higher parent ratings of repetitive behaviors on the RBS-R (total score) were significantly associated with parent SSS Impairment ratings ($r = 0.47$, $P = 0.021$), which is expected given the similarity of the behaviors being rated. Otherwise, parent ratings of autism symptoms (ASSQ, SRS), ADHD (CPRS, ADHD Rating Scale-IV), OCD (CYBOCS), and anxiety (MASC) were not significantly associated with any stereotypy ratings (all $P > 0.11$).

(1) Primary outcomes: changes in stereotypy severity

(a) Determined by phone call assessments: Primary outcomes in the ITT group, for data obtained by telephone, were assessed by comparing each individual's SSS Motor and SSS Impairment scores obtained at Call #1 to the last available assessment value (n = 24). Data, assessed using repeated measures ANOVAs; means, and standard deviations, are listed in Table 2. Compared with the first telephone call, there were significant reductions in SSS Motor and SSS Impairment scores (both $P < 0.001$). Calculated change ratios were −0.23 for SSS Motor and −0.31 for SSS Impairment scores. Table 3 shows the means and standard deviations for SSS Motor, SSS Impairment, and PGI-I scores at each of the telephone contacts. Results demonstrate a progressive improvement in all outcome measures. At the final telephone call assessment, there were no significant differences between scores for boys and girls on SSS Motor or PGI-I scores; however, boys had significantly higher ratings than girls on the SSS Impairment score ($P = 0.003$).

**TABLE 2.**
Intent-to Treat-Analysis: Comparison of Primary Outcome Measures at Baseline and Last Follow-Up Assessment (Calls)

| | Call #1 | | Last Post-DVD Call | | | | Change Ratio | |
|---|---|---|---|---|---|---|---|---|
| | (n = 24*) | | (n = 24) | | | | (n = 24) | |
| Measure | Mean | SD | Mean | SD | *P* | $\eta^2$ | Mean | SD |
| SSS Motor | 10.67 | 1.88 | 8.21 | 1.91 | <0.001 | 0.668 | −0.23 | 0.16 |
| SSS Impairment | 17.50 | 11.51 | 12.08 | 8.84 | 0.001 | 0.414 | −0.31 | 0.31 |

Abbreviation: SSS = Stereotypy Severity Scale.
*Note:* Change Ratio = mean of (follow-up score − baseline score)/baseline score.
* Participants with at least one telephone call.

(b) Determined by independent REDCap assessments: Primary outcomes in the ITT group, for data obtained via the internet (REDCap), were assessed by comparing each individual's SSS Motor, SSS Impairment, and SLAS scores obtained during the screening process to their last available assessment value (n = 22). Data, evaluated using repeated measures ANOVAs and means and standard deviations, are listed in Table 4. Compared with the baseline rating, there were significant reductions in all scores (all $P \leq 0.001$). Calculated change ratios were −0.30 for SSS Motor, −0.32 for SSS Impairment, and −0.54 for SLAS scores. Table 5 shows the means and standard deviations for SSS Motor, SSS Impairment, and PGI-I scores at each of the REDCap collection times. Similar to the telephone assessment, data showed gradually progressing improvement in all outcome measures. At final REDCap assessment, there were no significant differences between scores for boys and girls on any of the outcome variables.

(2) Secondary outcomes: changes in PGI-I scores

Descriptive statistics pertain to a subset of participants in the ITT group whose parents completed PGI-I assessments at call #2 (n = 23), call #3 (n = 23), and call #4 (n = 21). Participants were classified based on parent ratings of perceived change: *very much/much improved* (PGI-I score = 1-2), *improved* (PGI-I score = 3), *no change* (PGI score = 4), and *minimally worse/much worse* (PGI-I score = 5-6). Comparing the initial rating to final rating, the proportion of parent ratings for each category are as follows: *very much/much improved*—56.5%; *improved*—30%; *no change*—13%; and *minimally/much worse*—0.0%.

**TABLE 3.**
Parent Reports According to Length of Study Participation (Calls)

| | Call #1 | | Call #2 | | Call #3 | | Call #4 | |
|---|---|---|---|---|---|---|---|---|
| | n = 24 | | n = 23 | | n = 23 | | n = 21 | |
| Measure | Mean | SD | Mean | SD | M | SD | M | SD |
| SSS Motor | 10.67 | 1.88 | 10.17 | 2.10 | 9.00 | 2.43 | 8.14 | 1.98 |
| SSS Impairment | 17.50 | 11.51 | 16.09 | 10.33 | 13.91 | 9.41 | 11.90 | 8.73 |
| PGI-I | - | - | 3.52 | 0.51 | 3.00 | 0.52 | 2.19 | 0.87 |

Abbreviations: PGI-I = Parent Global Impression of Improvement; SSS = Stereotypy Severity Scale.

(3) Post-treatment questionnaire

Parents of 22 participants in the ITT group completed a post-treatment questionnaire. The majority of this group (n = 19) had provided follow-up assessments at Calls 2, 3, and 4, before finishing the post-treatment questionnaire. Participants (n = 19) indicated having watched the video several times (mean = 2.16 ± 0.83). All responders (n = 19) found the video "useful" and (100%) would "recommend it to others."

(4) Comparison of stereotypy ratings obtained by phone call versus REDCap

The last available assessment values for the SSS Motor and Impairment scores, obtained via phone call and via independent parent rating via REDCap (n = 22), were compared using paired samples *t* tests. For SSS Motor, the mean rating by phone call (8.18 ± 2.3) was slightly lower than the mean rating obtained via REDCap (8.55 ± 1.9), although the difference was not statistically significant ($P = 0.104$). For SSS Impairment, the rating by phone call (12.27 ± 8.7) was slightly higher than the rating in

**TABLE 4.**
Intent-to Treat-Analysis: Comparison of Primary Outcome Measures at Baseline and Last Follow-Up Assessment (REDCap)

| | Baseline Assessment | | Last Post-DVD Assessment | | | | Change Ratio | |
|---|---|---|---|---|---|---|---|---|
| | (n = 22*) | | (n = 22) | | | | (n = 22) | |
| Measure | Mean | SD | Mean | SD | *P* | $\eta^2$ | Mean | SD |
| SSS Motor | 12.27 | 1.39 | 8.55 | 2.28 | <0.001 | 0.764 | −0.30 | 0.18 |
| SSS Impairment | 15.45 | 7.39 | 10.45 | 7.85 | 0.013 | 0.262 | −0.32 | 0.43 |
| SLAS Score | 53.82 | 24.81 | 24.64 | 21.59 | <0.001 | 0.513 | −0.54 | 0.53 |

Abbreviations: SLAS = Stereotypy Linear Analog Scale; SSS = Stereotypy Severity Scale.
*Note:* Change Ratio = mean of (follow-up score − baseline score)/baseline score.
* Participants with at least one follow-up assessment provided by parents at month one or two.

**TABLE 5.**
Parent Reports of Stereotypy Severity According to Length of Study Participation (REDCap)

| | Baseline | | One Month | | Two Month | |
|---|---|---|---|---|---|---|
| | n = 38 | | n = 22 | | n = 21 | |
| Measure | Mean | SD | Mean | SD | Mean | SD |
| SSS Motor | 11.92 | 1.84 | 10.77 | 2.09 | 9.48 | 2.32 |
| SSS Impairment | 16.58 | 8.79 | 14.09 | 9.08 | 10.00 | 7.75 |
| SLAS Score | 52.39 | 24.48 | 30.68 | 19.48 | 24.38 | 22.09 |

Abbreviations: SLAS = Stereotypy Linear Analog Scale; SSS = Stereotypy Severity Scale.

REDCap (10.45 ± 7.9), but, again, the difference was not statistically significant ($P$ = 0.213).

## Discussion

Motor stereotypies in otherwise healthy children (primary CMS) have the potential to cause psychosocial issues or interfere with the performance of academic activities due to their frequent occurrence and prolonged duration. The value of behavioral therapy, in particular awareness training, and the differential reinforcement of other behaviors has been previously observed in 8 to 12-year-old children with therapy provided either in person by a therapist[6] or via a home-based, parent-directed program.[7] The current report describes the efficacy of a *combined* behavioral approach, i.e., home-based plus direct telephone contact with a behavioral therapist, in five to seven year-old children with primary CMS.

Our results suggest that a combined behavioral approach is beneficial, based on a reduction of scores from baseline to last assessment; documented by both telephone interviews and REDCap questionnaires. More specifically, results show a significant reduction in all primary outcome measures: SSS Motor (23% telephone and 30% REDCap), SSS Impairment (31% telephone and 32% REDCap), and SLAS (54% REDCap). These results compare very favorably to previous data obtained in 8-12-year-old children: (1) results from a three month parent-administered home-based treatment protocol showed REDCap reductions of SSS motor (15%), SSS Impairment (24%), and SLAS (20%)[7]; and (2) results from in-person behavioral psychologist administered therapy showed reductions of SSS motor (14%), SSS Impairment (33%), and SLAS (45%).[6] Evidence demonstrating the benefit of combined therapy also includes PGI-I and post study questionnaire results. In this study, the mean PGI-I value at call #3 (three weeks) was 3.0 and at call #4 (two months) 2.19 as compared with values in the DVD-only protocol[7] of 3.37 at one month and 3.03 at two months. Parent completed post-treatment reports showed that all responders found the therapy “useful” and would “recommend it to others.”

Although all prior behavioral studies in primary CMS have had slightly different formats and populations, the aforementioned data suggest that the best results were attained using the combined home-based plus therapist-assisted approach. In addition, this was the first study showing relatively similar beneficial outcomes in both the SSS Motor and SSS Impairment scales, suggesting that the combined treatment approach may have positive impact on both the movements themselves, *and* the functional impact of the condition. Having therapist involvement in the treatment protocol also improved subject retention. For example, in a prior home-based parent-administered treatment protocol, using the same instructional DVD but lacking contact with a therapist, there was a 30% loss of subjects between receipt of the DVD and a one month REDCap assessment and an additional 30% loss between one and two month REDCap assessments. In contrast, in the current study (between calls #1 and 4) there was only a 12.5% dropout rate.

An additional important aspect of any treatment protocol is the selection of a patient population that would be best suited for a particular therapy. In a prior report, it was suggested that parents who have children with a greater stereotypy burden would be more likely to participate in an active process that involved time, motivation, and compliance.[7] In the current report, however, severity of stereotypy was not a distinguishing factor in comparisons between the ITT and the LTF groups. In contrast, enrolled individuals with greater ADHD symptomatology, documented in the screening process, were more likely to drop out. Study results also identified a progressive reduction in SSS Motor and SSS Impairment scores over the two-month course of therapy. Hence, families who are willing to commit more sustained efforts toward home-based therapy are more likely to observe a greater beneficial response.

The precise pathophysiological mechanism for primary stereotypies is unknown with hypotheses including psychological,[16] a substitutive behavior for imaginative activities,[17] poor sensorimotor integration,[18] and a pathophysiological change in the habitual behavioral pathway (premotor and/or supplementary motor area to putamen).[4] The latter theory for primary CMS is supported by a volumetric magnetic resonance imaging study showing significant reduction in the putamen,[19] and a 7T magnetic resonance spectroscopy report displaying reduced gamma-aminobutyric acid (GABA) in the anterior cingulate and striatum.[20] Preliminary results in resting state functional magnetic resonance shows reduced connectivity between prefrontal and striatal regions, which may impose a secondary effect on top-down motor inhibitory control. Future studies, in children pre– and post–home-based-therapist-assisted behavioral treatment, would be of value in clarifying the anatomical location of stereotypic behaviors.

Despite the use of established stereotypy measurement scales[6,8] and the documentation of a reduction in severity, this study has several recognized limitations. First, although comparisons between stereotypy ratings obtained by telephone were comparable to those collected via the Internet, both were solely parent based. Hence, as emphasized by others,[7] future evaluations need to include objective confirmation by experienced investigators and/or by direct measurement of the movements themselves using actigraphy-type devices. Second, this protocol lacked a control comparison group, i.e., a therapist who provides only encouragement, but no helpful

suggestions, or a DVD that provides educational material but suggests ignoring the stereotypies. The investigators are also aware of studies suggesting that early-onset behavioral therapy is beneficial for conditions such as externalizing behavior problems,[21] and recognize the disparity between the age of onset of stereotypies in the study population (12.2 ± 10.1 months) and the patient age at the initiation of therapy (5.96 ± 0.81 years). Lastly, in retrospect, an additional telephone call between weeks three and eight would have permitted an opportunity to modify the DRO and possibly further enhance improvement.

In conclusion, there is no proven medication for the treatment of primary complex stereotypies whereas behavioral therapy has been shown to be beneficial. Based on the sequence of publications, the authors have recommended in-person therapy with a behavioral psychologist and the use of an instructional DVD for home-based parent-delivered behavioral therapy. Although only confirmed in a younger population, we would now advocate for the use of a combined home-based therapist-assisted therapy for all age groups. Future studies are required to address the issues of a randomly assigned control group, clinician-administered assessments, and the role of telemedicine.

The authors thank the Nesbitt-McMaster Foundation, the Klump and Graves families, and the many other donor families who have made this work possible through acts of philanthropy. This research was also supported by UL1 TR000424 and U54 079123. Special thanks to Lisa Ferenc, Kelsey Crowley, and Farhan Augustine for their assistance in manuscript preparation.

## Supplementary Data

Supplementary data related to this article can be found at https://doi.org/10.1016/j.pediatrneurol.2018.05.004.

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