Comparative Effects of Bilateral Hand Splints and an Elbow Orthosis on Stereotypic Hand Movements and Toy Play in Two Children With Rett Syndrome

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A single-subject rapidly alternating treatment design was used to compare the effectiveness of bilateral hand splints and an elbow orthosis in decreasing stereotypic hand behaviors and increasing toy play in 2 children with Rett syndrome. The subjects' responses were compared across three treatment conditions: no intervention, hand splints, and elbow orthosis. The order of the treatment phases was randomly selected for each subject. Data were collected in both a free-time condition and a toy-play condition; the outcome measures were stereotypic hand movements and hand-to-toy contact. Both subjects demonstrated a decrease in stereotypic hand movements and a corresponding increase in toy contact with the use of the elbow orthosis. The bilateral hand splints had no obvious treatment effect.

Rett syndrome is a neurological disorder of undetermined etiology, first described by Andreas Rett in 1966, but has just recently been recognized as a distinct clinical entity in this country (Hagberg, Aicardi, Dias, & Ramos, 1983). The syndrome has been reported only in females, with an incidence rate of approximately 1 in 10,000 (Trevathan & Adams, 1988). Rett syndrome is characterized by apparently normal development during the first 6 to 18 months of life, followed by developmental regression, loss of purposeful hand use with the appearance of stereotyped hand-washing movements, deceleration of head growth, gait apraxia and jerky truncal ataxia, autistic-like behaviors, and severe mental retardation (Hagberg, Goutieres, Hanefield, Rett, & Wilson, 1985).

The stereotypic hand movements and concurrent loss of purposeful hand skills are among the most striking characteristics of Rett syndrome and are necessary criteria for a positive diagnosis (The Rett Syndrome Diagnostic Criteria Work Group, 1988). The hand stereotypes may include hand-washing movements; hand wringing, licking, and sucking; the bringing of the hands to the mouth; and the stretching and flexing of the joints of the middle finger. Ataxia, sometimes accompanied by tremor, may also interfere with hand function. In addition, persons with Rett syndrome frequently do not demonstrate a pincer grasp (or any functional grasp) and may have abnormal movement patterns such as adduction of the thumbs and pronation of the forearms (Olsson & Rett, 1985). In some cases, self-injurious behavior, such as hand biting, is present.

Although the fine motor problems and stereotyped hand movements of Rett syndrome have been described in detail by several authors (Budden, 1986; Holm, 1985; Lieber, 1985), few have addressed the issue of treatment. Nomura, Segawa, and Hasegawa (1984) noted anecdotally that psychological means were unsuccessful in decreasing the hand movements and that medication was also ineffective. Iwata, Pace, Willis, Gamache, and Hyman (1986) used operant techniques to decrease hand biting in 2 subjects with Rett syndrome and to increase the toy play of 1 subject.

Several recent studies have investigated the effectiveness of upper extremity orthoses as a therapeutic technique. Naganuma and Billingsley (1988) used bilateral hand splints to greatly reduce stereotypic hand behaviors in 3 adolescents with Rett syndrome. One of the subjects also showed an increase in finger-feeding skills. However, in a replication study done by Tuten and Miedaner (1989) with two 5-year-old subjects, no change was found in the occurrence of stereotypic hand behavior with the application of identical hand splints, and no increase in purposeful hand use was observed.

Aron (1990) used elbow splints with 8 subjects ranging in age from 2 to 14 years. The splints prevented elbow flexion and hand-to-mouth movement. Using parental interview and clinical observation, Aron reported a de-
crease in hand-wringing behavior in all 8 subjects and increased hand use in 1 subject. Increased socialization and interaction with the environment were also reported. Three of the subjects wore the splints on only one arm. In a previous single-subject study (Sharpe & Ottenbacher, 1990), we investigated the effect of an elbow orthosis applied to the nondominant arm of a 5-year-old subject. In that study, the subject demonstrated modest improvement in finger-feeding skills while wearing the elbow splint.

The purpose of the present study was to compare the effectiveness of bilateral hand splints and an elbow splint in decreasing stereotypic hand behaviors and increasing toy play in 2 children with Rett syndrome. I investigated whether the use of these treatment techniques would suppress the hand stereotypes and whether suppressing them would lead to an increase in purposeful hand use.

Method

Subjects

Subject 1 was a 6-year-old girl whose condition was diagnosed as Rett syndrome at age 2½ years. She was attending a full-day functional living skills program in a public school. She was ambulatory and exhibited the stereotypic hand-wringing movements and hand-to-mouth behaviors characteristic of Rett syndrome. She was right-hand dominant and was able to grasp objects and finger feed independently.

Subject 2 was also 6 years old and was attending a program for multiply handicapped students in a public school. She was born 2 months premature and demonstrated a right hemiplegia that was probably secondary to prematurity. Her condition was diagnosed as Rett syndrome at age 5 years. Subject 2 was ambulatory and demonstrated moderate spasticity in her affected right upper extremity. Her hand stereotypes were primarily unilateral and included waving her left hand, mouthing it and sucking on her fingers, and wringing both hands. Subject 2 required complete assistance with all self-care skills and demonstrated only occasional spontaneous reaching for toys or food. It should be noted that under the diagnostic criteria for Rett syndrome that were recently developed by the Rett Syndrome Diagnostic Criteria Work Group (1988), this subject would not be considered to have a typical case of Rett syndrome due to her cerebral palsy.

Splints and Orthosis

The elbow orthosis was fabricated out of a lightweight plastic material (needlepoint canvas), covered with vinyl, and secured with touch-fastener straps. It permitted only a few degrees of elbow flexion (see Figure 1). This design was based on a commercially available model, however, other types of elbow orthoses, such as a dropout splint, which allows more active movement, would probably be equally effective. Subject 1 wore the orthosis on her nondominant left arm. Subject 2 wore the orthosis on her dominant left arm. Preliminary observations of Subject 2 indicated that when she worked with the orthosis on her nondominant arm, little change was noted in stereotypic hand movements because these movements were almost exclusively unilateral. By applying the orthosis to her unininvolved left arm, I hoped to interrupt the hand movements and encourage more active movement with the right upper extremity.

Thumb abduction splints were fabricated according to the specifications in the Naganuma and Billingsley study (1988). The splints positioned the subjects’ thumbs in abduction but did not restrict functional wrist or finger movement. The splints were individually fitted to each child.

Procedure

A single-subject rapidly alternating treatment design, as described by Ottenbacher (1986), was used to compare the subjects’ responses in three treatment conditions: no
intervention (A phase), bilateral hand splints (B phase), and elbow orthosis (C phase). The order of the treatment phases was randomly selected for each subject. This randomization reduced the possibility of order effects, a potential limitation of the alternating treatment design (Ottenbacher, 1986). Data were collected each day in both a free-time condition and a toy-play condition, with the subject wearing either the splints or the elbow orthosis or neither.

The study was carried out in a quiet area of each child's classroom. The subject was seated on a rug on the floor with the classroom aide seated nearby to record data. The orthosis or splints were put on immediately before each session.

Each session consisted of 5 min of free time (no toys available), followed by 5 min of a toy-play condition in which three toys were positioned within the child's reach. The classroom aide verbally encouraged the subjects to play with the toys but did not prompt them physically. The toys were chosen each day from the selection available in the classroom. They were varied from day to day to avoid boredom, but each subject's particular favorites were regularly included. If the subject threw a toy or moved it out of reach, the aide returned it to the original position on the rug.

Treatment sessions were held at approximately the same time each day, 5 days per week, although school vacations and illness intervened. A total of 30 sessions was conducted for each child. Following the conclusion of the alternating treatment phase of the study, five follow-up sessions were conducted. No attempt was made to coordinate the sessions between the 2 subjects. This study was conducted as two separate single-subject alternating treatment designs.

Data Collection

Outcome measures were stereotypic hand movements and hand-to-toy contact. During each 5-min free-time period, data were collected on the subject's stereotyped behaviors with the use of a 10-sec time sample. At the end of each 10 sec, the observer recorded whether the subject was engaging in stereotyped hand behaviors. The recording continued at 10-sec intervals for the entire 5 min (a total of 30 intervals). For purposes of data collection, stereotyped hand movements were defined as any repetitive, nonpurposeful contact of one hand with the other (including wringing, clapping, washing movements, and pulling on fingers and thumbs), one or both hands in contact with the mouth, or unilateral hand movements such as waving the fingers or rubbing the fingers against the palm.

In the toy-play condition, the 10-sec time sample was also used. At the end of each 10 sec, the observer recorded whether the subject was engaging in toy play, which was defined as having one or both hands in contact with a toy. Stereotypic behaviors were also recorded during this 5-min session, and it was possible that both behaviors might occur simultaneously. Both behaviors were recorded as occurring or not occurring at each 10-sec interval. No attempt was made to record duration or frequency of a behavior within an interval.

Reliability

Point-to-point interrater reliability was obtained for both stereotypic hand movements and toy play by having an independent observer record data across 30% of all three conditions. An agreement was scored when both observers recorded that the behavior occurred or did not occur during the time sample. A disagreement was scored when one observer recorded occurrence and the other recorded nonoccurrence. I calculated reliability by dividing the number of agreements by the number of agreements plus disagreements and multiplying by 100 (Kazdin, 1982). Interrater reliability for Subject 1 ranged from 80% to 99% across all phases, with a mean of 92%. For Subject 2, the mean reliability was 93%, with a range of 84% to 99%. There was no significant difference in the number of agreements across the two different outcome measures.

Data Analysis

The results were analyzed by graphic presentation and visual inspection of the data. Measures of percentage overlap were computed for data from the various phases.

Results

Stereotypic Hand Movements

To facilitate comparison of performance, I collapsed the data across phases. Subject 1 showed fairly wide variability across all three phases in the free-time condition. However, the mean number of stereotypic hand movements in the A phase (no intervention) was 22.5. In the B phase (splints), the mean number of stereotypic hand movements was 21.9, and in the C phase (elbow orthosis), 16.4, indicating that the stereotypic behaviors occurred less frequently when the subject was wearing the orthosis. Calculation of percentage overlap shows that 100% of data points in the B phase overlapped into the range of the no intervention (A) phase, while only 60% of the data points in the C phase were overlapping with Phase A.

The data for Subject 2 in the free-time condition showed a more dramatic difference in stereotypic hand behaviors between the C phase (orthosis) and the A and B phases. This might be expected because Subject 2's hand movements were primarily unilateral and the orthosis was applied to her dominant arm. The mean number of hand movements for this subject in the baseline phase was 27.1; in the splint phase, 22.1; and in the orthosis...
of the data points in the B phase overlapped into the range of the A phase. Eighty percent of the data points in the B phase overlapped into the A phase. In the follow-up phase, the data for both subjects were consistent with that of the first A phase.

Data for stereotypic hand movements of both subjects in the toy-play condition with data points collapsed for each phase were analyzed. In the toy-play condition, Subject 1 showed a significant reduction in stereotypic hand movements when she was wearing the orthosis (m = 7.5), as compared to the A phase (m = 21.5) and the B phase (m = 19). None of the data points in the C phase were in the range of the A phase, while 90% of data points in the B phase overlapped into the A phase.

The data for Subject 2 in the toy-play condition showed a similar pattern. The mean number of stereotypic hand movements in both the A phase (no intervention) and the B phase (splints) was 25.6. In the C phase (orthosis), the number decreased to 12.3. All of the data points in the B phase overlapped into the A phase, while only 40% of the data points in the C phase were in the range of the A phase. In the follow-up phase, all data points for each subject were within the range of their first no-intervention phase.

**Toy Play**

The number of times that each subject touched a toy with the data points collapsed for each phase was analyzed. Subject 1 showed almost twice as much hand-to-toy contact in the C phase (m = 19.9) as she did in the A phase (m = 10.1). The mean number of toy contacts in the B phase (12.5) was only slightly above that in the A phase. Eighty percent of the data points in the B phase overlapped into the range of the A phase, while only 30% of the data points in the C phase were within that range. In the follow-up phase, the mean number of hand-to-toy contacts was 11.8, which is consistent with Subject 1's performance in the first no-intervention (A) phase.

Subject 2 also showed significantly increased toy contact while she was wearing the elbow orthosis. It is interesting to note that most of the contacts were made with her restrained dominant left hand. In the no intervention (A) phase, she touched a toy on only 2 of the 10 days (mean number of contacts = 0.3). In the splint phase (B), toy contact was made on only one of the 10 days (m = 1.1). While wearing the orthosis (C), Subject 2 made contact with a toy on 8 out of 10 days, with a mean of three hand-to-toy contacts. Percentage overlap was not calculated for these data because the responses occurred so infrequently in the no-intervention (A) phase. In the follow-up phase, Subject 2 made contact with a toy on 2 of the 5 days.

In Figure 2, hand-to-toy contact and stereotypic hand movements for Subject 1 in the toy-play condition are represented (with data points collapsed separately for each phase), so that the relationship between the two outcome measures can be examined. It can be clearly seen that as stereotypic hand movements decrease, hand-to-toy contacts tend to increase. This is particularly evident in the C phase (orthosis), where the relationship between the two outcome measures is reversed, indicating that hand-to-toy contact was occurring more frequently than the stereotypic hand behaviors. In Figure 3, the same comparison is made for Subject 2, and a similar pattern is observed.

**Discussion**

In both subjects, a decrease in stereotypic hand behaviors occurred with the use of the elbow orthosis. This decrease was observed in both the free-time and the toy-play conditions. The data also show that with the reduction in stereotypic hand movements, there was a corresponding increase in the number of hand-to-toy contacts. Application of the bilateral hand splints had no treatment effect on either stereotypic hand movements or toy play. There was no evidence of maintained effects of wearing the orthosis. The data in the follow-up phase, which served as a second baseline condition, were consistent with the no-intervention results in the alternating treatment phase of the study.

Although both subjects showed measurable changes in target behaviors with the use of the elbow orthosis, their responses differed. Subject 1 (who had functional hand use) demonstrated a greater reduction in stereotypic hand movements in the toy-play condition than in the free-time condition with the orthosis. No significant differences in the number of stereotypic hand movements were observed in either the baseline or the splint phases. One can hypothesize that the toys were an interesting stimulus for her and that the orthosis enabled her to interact with them more freely; consequently, she spent less time engaging in stereotypic hand movements. Subject 1 played with the toys by grasping, mouthing, and frequently throwing them. During the free-time condition, this subject often seemed bored and sought adult attention.

Subject 2, who had no functional hand use, showed a consistent reduction in stereotypic hand movements across both the toy-play and the free-time conditions with the use of the orthosis. When she made contact with a toy, it was usually by means of placing her open left hand (the restrained arm) on the toy in what often appeared to be a random movement. However, the increase in the number of hand-to-toy contacts when she was wearing the elbow orthosis suggests that her movements were not as random as they appeared.

The results of this study are consistent with the findings of Aron (1990), who observed decreased hand stereotypies in 8 subjects who wore elbow orthoses. However, only one of her subjects demonstrated a
30 A Phase B Phase C Phase

Figure 2. Comparison of stereotypic hand movements with number of hand-to-toy contacts for Subject 1. Note. A phase = no intervention; B phase = splints; C phase = elbow orthosis.

corresponding increase in hand use with the orthoses. The findings are also consistent with Sharpe and Ottenbacher’s (1990) single-subject study in which modest improvement in finger-feeding skills was evident with the use of the elbow orthosis.

The results of this study are not consistent with Naganuma and Billingsley’s (1988) finding that application of bilateral hand splints reduced stereotypic hand movements in 3 subjects with Rett syndrome. Although the splints used in Naganuma and Billingsley’s study and the present study were similar, the splint application schedule was different. The subjects of the Naganuma and Billingsley study wore the hand splints daily, the entire time they were at school, whereas in the present study, the splints were applied immediately before each session. This may have reduced the splints’ effectiveness in interrupting the stereotypic behaviors. However, Tuten and Miedaner (1989), in their replication study, also observed no changes in either stereotypic hand behaviors or functional hand use when their subjects wore bilateral hand splints. The subjects in both the replication study and in the present study were younger than the subjects in the Naganuma and Billingsley study, which may have affected the outcome.

The relatively large treatment effect produced by the use of the elbow orthosis in the subjects’ natural classroom setting suggests that it may be a practical and effective means of decreasing stereotypic hand movements and increasing purposeful hand use in some persons with Rett syndrome. Naganuma and Billingsley (1988) theorized that the stereotypic hand movements of Rett syndrome may function as a competing behavior to purposeful hand use and that by decreasing these behaviors, development of more functional skills can be encouraged. The elbow orthosis interrupts the hand stereotypes, thus giving the wearers an opportunity to use their hands to explore and interact with their environment. The extent to which they are able to do so may depend on their level of cognitive ability.

The elbow orthosis is easy to fabricate, inexpensive, and simple to apply. However, there are limitations to its effectiveness. One major drawback is that bilateral hand use is precluded when the orthosis is worn. Another limitation is that although the orthosis effectively interrupts hand-to-mouth behaviors, hand wringing is not eliminated, although it occurs less frequently. Other possible disadvantages of use of the elbow orthosis include the possibility of frustration and discomfort for the wearer due to being restrained, although this was not observed with either of the subjects in the present study. The possibility
also exists that the wearer might exhibit an increase in the intensity of the stereotypic hand movements immediately after the orthosis is removed (this also was not observed in the present study).

The elbow orthosis is not an appropriate treatment technique for use in all situations. For example, it would be of limited practical use with Subject 2 in teaching feeding skills, because in addition to interrupting hand stereotypies, it also made it impossible for her to bring her dominant hand to her mouth. Subject 1 was a better candidate because when her nondominant hand was restrained, she used her dominant hand more frequently, and indeed she continues to wear the orthosis during mealtimes both at home and at school.

Implications and Conclusion

There are many implications for further research in this area. For example, it is necessary to explore the effectiveness of the elbow orthosis across other conditions and other subjects of various ages and skill levels. It would also be interesting to investigate whether the frequency of stereotypic hand behaviors increases from baseline levels immediately after the orthosis is removed and whether the amount of time the orthosis is worn affects its effectiveness in reducing hand stereotypes. Comparative studies are also needed to further explore the relative effectiveness of the bilateral hand splints and the elbow orthosis.

To date, only a handful of investigators have addressed the issue of management of stereotypic hand movements in Rett syndrome. In the present study, I used an alternating treatment design to compare two treatment techniques—bilateral hand splints and an elbow orthosis. I found that stereotypic hand movements were reduced and toy contact increased with the use of the elbow orthosis in 2 subjects with Rett syndrome. In contrast to previous findings by Naganuma and Billingsley (1988), bilateral hand splints had no obvious treatment effect. Additional research is needed to further explore the efficacy of these and other treatment techniques so that the results can be generalized to the larger population of persons with Rett syndrome.

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References


