The effect of hand splints on stereotypic hand behavior in Rett's syndrome

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The purpose of this study was to examine the effect of hand splints and one elbow restraint on persistent stereotypic hand movements of four girls with Rett's syndrome.

Among the most characteristic features of Rett's syndrome are stereotypic hand wringing and loss of previously acquired functional hand skills. Hand splints and one elbow restraint were used in this study. The subject's stereotypic hand behavior and functional hand use were calculated from five-minute segmental video tape recordings. The study consisted of three phases: baseline, intervention, and withdrawal.

All subjects demonstrated a decrease in stereotypic hand behavior after the application of hand splints. Although splints showed a positive effect on hand movements in Rett's syndrome, they could also lead to other, undesirable, movements. Whether splints have a positive effect on the functional use of the hand should be investigated in more subjects.

Key words: Rett's syndrome, stereotypic movements, hand splints.

Rett's syndrome is a progressive encephalopathy in females that appears during the first 18 months of life. It was described in Vienna in 1966 by Andreas Rett who noticed a similar appearance and complex wringing motions in the hands of several of his female patients[1,2]. Rett's syndrome is a neurological disorder affecting predominantly females and characterized by regression, and loss of speech and purposeful hand use after six to 18 months of almost normal development. Postnatal microcephaly, hand dyspraxia, stereotypic hand movements, ataxia, severe mental retardation and abnormal breathing are among the most characteristic features[3-8]. Naganuma and Billingsley[9] reported that bilateral hand splints significantly decreased stereotypic hand behavior in three cases with Rett's syndrome and increased finger-feeding skills in one of the cases. Sharpe and Ottenbacher[10] used an elbow restraint splint to improve the functional hand use in a child with Rett's syndrome, and stated only minimal support for its use in improving finger-feeding skills.

Aron[11] used elbow splint in eight children with Rett's syndrome in 1990, and studied the self-injuring behavior of children with their hands. Results were very positive and indicated an increased socialization and interaction with the environment, and decreased hand-mouth movements and hand wringing behavior.

The aims of our study were to investigate the effects of hand splints and one elbow restraint in preventing seterotypical hand movements and to determine functional hand use in four cases.

Material and Methods

Four subjects who were diagnosed at the Pediatric Neurology Department of Hacettepe University Hospital were assessed. All of them were from Turkey. They fulfilled the Rett's syndrome diagnosis criteria. Pregnancy, birth and psychomotor development during the first year of life were normal. The mean age at onset was 14 ± 10.95 months. Mental retardation, stereotypic hand movements and loss of

purposeful manual skills were noted in all cases. All children's parents were positive regarding splint application.

Subject 1 was a nine-year-old girl whose stereotypic hand behaviors began at 12 months of age. Before the study, her hand behaviors consisted of pill-rolling, hand-to-mouth, squeezing and wringing. Her previous levels of functional hand use were very limited.

Subject 2 was a 10-year-old girl who began to show seterotypic hand behavior at about 2.5 years of age. At the start of this study, her hand movements consisted of hand wringing, squeezing, and hand-to-mouth; grinding of the teeth was also present. Before the intervention, she could finger-feed herself independently and drink a from a cup.

Subject 3 was an 11-year-old girl who began to show seterotypic hand movements at about eight months of age. Before this study, her hand movements consisted of hand-to-mouth, pill rolling, wringing, and squeezing; trunk swinging and teeth grinding were also present. She was successful in her functional hand movements.

Subject 4 was an eight-year-old girl who began to show stereotypic hand behavior at about six months of age. Before this study, her hand behaviors consisted of hand clapping, hand-to-mouth, squeezing and hand wringing; trunk swinging was also present. On examination of hand function, she was able to hold an object (biscuit) with help, but would drop it in one or two seconds; failing to eat it. Characteristics of subjects are given in Table I.

patient needed a pair of elbow splints to avoid self injury. These restraints permitted only a few degrees of elbow flexion (Fig. 2). Application of splints was gradually increased to five hours per day.

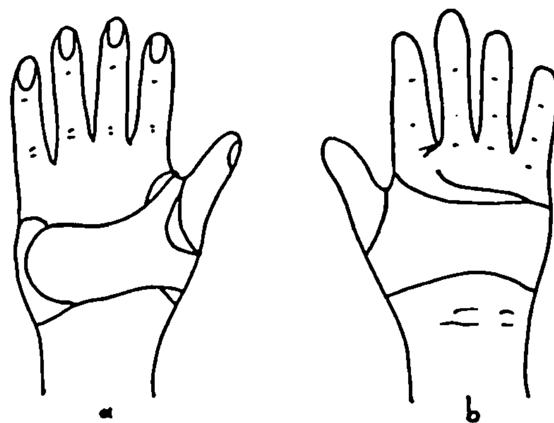

Fig. 1. Hand splint design a) dorsal view. b) palmar view.

Fig. 2. Elbow restraint.

Table I. Characteristics of Subjects With Rett's Syndrome

Subjects	Age (years)	Age at onset (months)	Stereotypic behaviors before splints
1	9	12	Hand-to-mouth, pill rolling, wringing, squeezing
2	10	30	Hand-to-mouth, squeezing, wringing, tooth grinding
3	11	8	Hand-to-mouth, pill rolling, wringing, squeezing, trunk swinging, tooth grinding
4	8	6	Hand-to-mouth, pill rolling, clapping, squeezing, wringing, trunk swinging

Hand splints and elbow restraints were made at Hacettepe University School of Physical Therapy and Rehabilitation. The splints were fabricated of plastozoid materials. The hand splints resembled a cuff circling each palm and positioned the thumb out from the palm into abduction (Fig. 1). Three elbow restraints were constructed for nondominant arms but one

Procedure

Stereotypical hand behavior was defined as: 1) hands in contact with each other and moving in any way (washing, wringing or clapping); 2) hands not in contact but one or both hands engaged in repetitive squeezing, pill rolling, or writhing or hand-to-ear movements; 3) one or both hands in contact with the lips or tongue

aside from when self-feeding. In addition, anterior-posterior and lateral trunk movements and teeth grinding were observed. Stereotypical hand behaviors were scored separately, totaling number of each stereotypical hand movement observed.

Functional hand use was defined as the subject's hand successfully holding a cracker and moving it to her mouth. The definition excluded times when the subject held food but failed to bring it toward her mouth for more than two seconds. As long as the subject had one hand in the process of grasping a piece of food, and bringing it to her mouth or placing it in her mouth, functional hand use would be scored even if the other hand were engaged in stereotypic hand movements. For the functional use of the hands, every touch of the hand on the cracker was considered, and the number of successful results was scored. Successful finger feeding was defined as the patient's successfully picking up a cracker and putting it in her mouth. If the piece of cracker fell out of her mouth as she was chewing, it was still counted as a success. If a piece fell as the subject was holding it, was counted as a failure. Food which was carried to a patient's lips successfully but fell before entering her mouth was also counted as a failure. In addition, grasping and releasing movements were assessed.

The study consisted of three phases: baseline, intervention and withdrawal. During the baseline phase, subjects did not wear hand splints or elbow restraints, and could initiate functional hand use or perform stereotypical movements as they preferred while in the occupational therapy unit. During the intervention phase, subjects wore splints on both hands for increasingly longer periods of time. As during the baseline phase, hand behaviors (stereotypic or functional) were not interrupted nor was any attempt made to reinforce them during the intervention phase. A brief withdrawal phase (return to baseline condition) was conducted in all subjects at the completion of the study to identify any maintenance effects of the hand splints. Subjects did not wear their hand splints during the 3rd phase, and could perform whatever hand behavior they preferred. In keeping multiple-baseline requirements, hand splints were applied in a sequential fashion to each subject only after stability in a behavioral level or trend was established. After 10 days of the baseline phase, subjects began to wear their elbow restraints. Application was gradually increased to 10 days, after which subjects began to wear their hand splints. The subjects wore the splints for a month. In the withdrawal phase, subjects did not wear hand splints or elbow restraints.

Data Collection

Videotape recordings were made during the entire study: for 10 days of baseline phase, during the intervention phase of 10 days when only elbow restraints were used, during the second part of the intervention phase of 30 days when both elbow restraints and hand splints were used, and lastly during the withdrawal phase when both elbow restraints and hand splints were removed. During these recordings, subjects were positioned so that both of their hands were placed on the table. The number of stereotypical movements was obtained from the five minute video recordings made during the free time just before the assessment of functional hand use. During these recordings, subjects who permitted headphones listened to music. In the following five minutes, functional hand use was evaluated regarding ability to eat a cracker placed on the table. Thus, each time the camera obtained a 5 minute recording of both hands for each patient.

Results

In the first subject, the hand wringing movement disappeared with the use of the elbow restraint, but movements were observed to increase during the withdrawal phase. Hand squeezing appeared following the use of the splint and restraint, with a greater performance on the left hand. Pill rolling increased on the right with the elbow restraint, but on the left with the elbow restraint, it disappeared. However, with the use of splint and restraint, it appeared again, and at the with drawal phase, it continued at an increasing rate. Bringing of the left hand to mouth disappeared with both elbow restraint and hand splint, but the movement was observed to increase during withdrawal phase. The hand wringing movement appeared on the right with the use of elbow restraint and hand splints.

In the second subject, hand wringing movement decreased with the use of restraint, but with hand splint, it increased. The right hand

squeezing movement decreased during the intervention phase. The left hand squeezing movement when compared with the right decreased more at intervention phase; however, at withdrawal phase, it returned to that observed at the beginning phase. The movement of the right hand to the mouth decreased following the application of the elbow restraint. It decreased further with the application of restraint and splint, but returned to level observed at beginning phase during withdrawal phase. The bilateral hand-to-mouth movement disappeared during the intervention phase except on the 15th day, but it appeared again during the withdrawal phase.

In the third subject, pill rolling decreased at a greater rate on left and right sides during the application of elbow restraint in the intervention phase, and this decrease continued during withdrawal phase too. Bringing of the hand to mouth increased on the right with the application of the elbow restraint. The hand wringing movement on the right appeared during intervention, and continued at the same level during the withdrawal phase.

In the fourth subject, hand wringing movement disappeared during intervention and withdrawal phases. Pill rolling disappeared with the application of elbow restraint but it appeared again at withdrawal phase. Bringing of the hand to mouth disappeared on the left, and did not reappear during the withdrawal phase. Bringing of the hand to mouth decreased on the right during the withdrawal phase.

Discussion

In children with Rett's syndrome, many complicated pictures appear with a variety of clinical findings[12-17]. Hard work is necessary to investigate the effects of different rehabilitation approaches[18,21]. When considering the cases in our study, although stereotypical hand movements were observed in all, the most frequent movements observed were hand-to-mouth movement, unilateral hand squeezing and wringing motion. In addition, trunk movement was observed in two children and tooth grinding in one. The children did not show any significant reaction to splinting except unruliness on the first day. The elbow restraint applied on the nondominant side prevented hand-to-mouth movement on that side; however, withdrawal of the restraint resulted in a return to levels observed during initial stage. The hand wringing motion observed in four children decreased with splints in three children, but only one of them had a decrease at the withdrawal phase. Again, in three children the hand squeezing movement decreased with splinting. This decrease continued during the withdrawal stage. The pill rolling movement appearing in three children showed a decrease in two children. On withdrawal of the splint, the decrease continued with one child. Studies using the elbow restraint or the hand splints show a decrease in the stereotypical movements[1,9,10,11,20,21].

Based on our observations, the application of splints in general led to somewhat of a decrease in stereotypic movements. The decrease appeared with the hand-to-mouth motion unilaterally. Trunk movement was observed in two children. Following the application of the splint, this movement decreased in one child. This may be attributed to the synergic movement with the application of the splint. Tooth grinding was absent in two cases before the application of splints but it appeared after wards. This may reflect the children's reaction to the splingt. With the splint application a decrease was observed in other movements, too, but with the removal of the splint, many cases returned to levels observed during the initial stage. This may have been due to the withdrawal of the splint itself, to the short period of splint application. In further studies, more objective results could be obtained by extending the period of splint application. In addition, by separate application of splints, their effects could be compared.

Feeding activity and the functional use of the hand also showed an increase with splint application in one child. This continued during withdrawal phase. Stereotypical hand movements in this child showed a decrease. This result shows that the decrease in the stereotypical hand movements may results in an independence in daily living activities. The application of elbow restraint to the nondominant side also resulted in a decrease in stereotypical hand movements on the dominant side. The hand splints used on the dominant side produced a reflex-inhibitory posture, with the stabilization of these two effects. This was also found to be positive in increasing independence. Whether or not splints

have a positive effect on the function use of the hand should be investigated in more subjects. In conclusion, although splints showed a positive effect on the hand movements of children with Rett's syndrome, it can also lead to other, undesirable, movements. For this reason, the effect of the splints in decreasing the movements should be shown more precisely with many more subjects and by examining the causes bringing about undesirable effects. The use of other occupational therapy approaches together with the effect of the splints should also be investigated. Our researches on these subjects continues.

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