CASE REPORT

Use of Splints to Develop Hand Skills in a Woman With Rett Syndrome

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Key Words: eating aids • occupational therapy (treatment) • stereotyped behavior

Rett syndrome challenges the occupational therapist because of the affected person's loss of or limited hand function, fear of movement, slow responses, abnormal tone, and stereotypic hand movements. In this case report, wrist and finger splints were fabricated for a 20-year-old woman with Rett syndrome who had lost all active hand control as a result of this disorder. After 3 years of use and one splint revision, grasp, pinch, and release have begun to emerge.

Background

A congenital, neurological disorder first described in 1966 by Andreas Rett, a neurologist from Vienna, Rett syndrome has only recently become known in the United States (Lindberg, 1991; Schleichkorn, 1987a; Sharpe & Ottenbacher, 1990). In 1983, Rett syndrome was described in an American neurology journal, and interest in it increased. Its incidence is believed to be 1 in 10,000 female births (Lindberg, 1991). Though it was thought to be a syndrome affecting females only, recently, two cases of males affected similarly were reported (Philippart, 1990).

From birth until 18 months of age, the child with Rett syndrome seems to develop normally, though delays in hand development can be noted early on (Nomura & Segawa, 1990). Initial symptoms, which begin to manifest between 6 and 18 months of age, are hand patting; excessive licking of toys or fingers; teeth grinding; protruding tongue; screaming; sudden, meaningless laughing; a general lack of attention to surroundings; and deceleration of skull growth (Lindberg, 1991; Nomura & Segawa, 1990). Stereotypic hand movements occur between 8 months and 3 years of age, and at that time, the child loses purposeful use of the hands. These movements include rubbing or wringing of the hands at the midline in front of the chest or the mouth and are the primary symptoms of Rett syndrome (Lindberg, 1991; Nomura & Segawa, 1990). Additional manifestations are breath holding and hyperventilation (Lugaresi, Cirignotta, & Montagna, 1985), tremor, hypotonia with spasticity developing with age, ataxia, chewing and swallowing difficulties, autistic-like behavior, and an inability to speak. As the disease progresses, the child frequently develops orthopedic complications (Hennessy & Haas, 1988). During the school years, seizures can occur as well. As the child leaves the teen years, gross motor losses accelerate, and most persons become wheelchair bound due to weakness, spasticity, and scoliosis (Lindberg, 1991).

Three studies have described the use of elbow or hand splints in the treatment of Rett syndrome (Aron, 1990; Naganuma & Billingsley, 1988; Sharpe & Ottenbacher, 1990). The results included reduced self-injurious behavior (Aron, 1990), decreased hand wringing (Aron, 1990; Naganuma & Billingsley, 1988; Sharpe & Ottenbacher, 1990), a positive effect on socialization, and in-
creased frequency of existing grasp and release (Aron, 1990). Increased duration of finger feeding and decreased amount of time required to eat have also been described (Sharpe & Ottenbacher, 1990). In one study, when splints were removed, target behaviors returned to baseline level (Naganuma & Billingsley, 1988).

Case History
This case report describes the occupational therapy treatment of a young woman who was diagnosed at the age of 15 years as having Rett syndrome. It needs to be understood that at the time of her diagnosis in 1986, information about the progression of the disease, its symptoms, and intervention strategies were sparse. Therefore, the development of her occupational therapy program had to be based solely on observations of this woman’s interests and physical needs. The development of this strategy is described below, with a focus on splinting.

This woman’s childhood history appeared to be a normal one and included sitting at 6 months of age, babbling, displaying social behavior, crawling, and walking independently at 11 months of age. However, speech never developed, and at 20 months of age, she began to withdraw. Her parents suspected a hearing problem and initiated several evaluations at noted hospitals. By the age of 3 years, she had lost the ability to use her hands, her gait was awkward, and spasticity had developed. In 1986, she was diagnosed as having Rett syndrome.

I started to work with her when she was 15 years old. At that time, this young woman had excessive flexor spasticity in her upper extremities, extensor spasticity in her lower extremities, tremor, contractures in the right wrist and left forearm, kyphoscoliosis, no trunk rotation, facial grimacing, breath holding, hyperventilation, drooling, poor bite, and no speech. She held her hands under her chin most of the time, with her fingers and wrists flexed and her left forearm in pronation (see Figure 1). Her symptoms were atypical in that she did not have a hand-wringer pattern of movement. Hypermobility was present at all joints of the fingers and thumbs of both hands. Occasionally, she would pat her lap tray, run her right hand across her forehead, or suck on her left fingers, but had no active grasp or release. She could not place her hands in her lap or keep her hands down if placed there.

At age 15 years, this young woman was a member of a public school class of students who had special physical and cognitive needs. Occupational, physical, speech, and music therapies were available to the students when appropriate and were integrated into the educational program.

Management of the Splinting Program
In her class at school, this student spent her time with her hands under her chin, as an observer. Because she could not place her hands on her switch plate, she was provided with a switch plate to enable her to operate various machines, such as a tape recorder. Other than her sporadic switch use, she was dependent on those about her for all of her needs, requiring hands-on assistance in art class, in eating, and in any other school-related activity. When trying to encourage her to keep her hands on her switch plate, I noticed that if her wrists were stabilized in a neutral position, she could keep her hands on the plate for several seconds, rather than momentarily. This meant that, brief as it was, the tendency to pull the hands up under the chin was reduced. In feeding, if the right wrist was stabilized, she could approximate a crude grasp on her spoon (though it appeared to be tenodesis action). This behavior was observed both by myself and by the classroom aide who assisted me with feeding.

As a result of these observations, simple dorsal wrist splints were fabricated and applied. Five months later, gains included the client’s ability to keep her hands in her lap with the splints on for 20 sec and the ability to hold onto her spoon and get it to her face 8 out of 10 times. (For feeding, the student used a deltoid suspension sling as well as a scoop dish and built-up handled spoon, angled for her needs.) She could not aim accurately for her mouth.

Figure 1. The student with Rett syndrome without splints. Note that stereotypic hand posturing is present.
One year later, the student still had no active grasp or release, but on two occasions had actively opened her hand with splints on at the appropriate time to receive her spoon. She was tolerating hands in lap with splints on for 5 to 30 min. When eating, she was holding the spoon for each bite and bringing it to her mouth each time, needing help with aiming 75% of the time. Occasionally, she was attempting to bring the spoon back to the plate. In addition, on one occasion I observed the student reaching for a favorite cookie, with the thumb and index finger opening slightly as if to grasp it. This emerging ability had not been observed prior to this and was not repeated. Realizing that the potential for active grasp as well as pinch and release was present, I decided to fabricate another set of splints to position the hand for grasp.

The splints were revised to include an opponens component, which secured the thumb in abduction (see Figure 2). Volar finger gutter splints were also fabricated to stabilize the interphalangeal joints of the right index finger as well as the thumb (see Figure 3).

Thirty-four months after the first splint application, this student (using splints and the suspension system) could bring her spoon to her mouth independently with 50% accuracy, needing help with her aim. She was able to return her spoon to her plate 40% of the time, but could not scoop. In finger feeding, she was able to open the thumb and index finger and reach for favorite snacks 1 out of 16 times (see Figure 4). She was able to close her fingers onto the food, but could not grade her pinch pressure or oppose the two finger pads with accuracy. She also had difficulty maintaining her pinch as she moved the hand toward her mouth.

With wrist and finger splints applied, she was now able to keep her hands in her lap for 45-min periods (see Figure 5). At the end of the school year, the student was observed fingering a textured ball for the first time in gym. This was her first attempt at exploring her environment with her hands since babyhood. At about the same time, she was observed sitting in her chair with both hands in her lap with no splints on. No one had positioned her that way; she had been able to place her hands in her lap independently.

Although the focus of this report has been on feeding, gains have been noted in hand function in other areas of the student’s school day by her teachers, classroom aides, and therapists. These areas are gym, art, cooking, music, arts and crafts, and participation in vocational and community tasks. There has been a noticeable increase in the student’s sociability as well.

Several other aspects of this student’s occupational therapy program need to be mentioned to understand the success achieved with splinting. From the time she was 15 years old, she has received sensorimotor treatment several times a week in occupational therapy. Recently, myofascial release has been incorporated with good results. This emphasis on tone reduction and facilitation of more normal movement reactions has given her greater freedom to move (Hanks, 1990). Without sensorimotor treatment, it is unlikely that she would have...
The student could feed herself with self-feeding splints applied with the use of an overhead deltoid suspension system. Achieved as much mobility. Maintaining passive range has been a priority, as she has abnormal tone and contractures. She is given plenty of time to respond, a necessary component when working with persons with Rett syndrome, as they can be dyspraxic and need time to sort out how to accomplish a task (Hanks, 1990; Kjoerholt & Salthammer, 1990; Lindberg, 1991).

Discussion

In this case, stereotypic hand behaviors were reduced through the application of dorsal wrist splints. As a result, this 20-year-old student was able to keep her hands in her lap and, over time, hand grasp, pinch, and release emerged. The application of finger splints gave her a mechanism to allow her to use her fingers functionally. It was interesting that given this mechanism, the student seemed to understand how to use those fingers even though she had not done so in 17 years.

It is not understood why the repeated daily application of wrist splints in school reduced this student's stereotypic hand behaviors (e.g., constant positioning of fisted hands under her chin). Was it the elimination of one component of her flexion synergy? Was wrist flexion a trigger mechanism that called for a total flexion reaction of the limb? Was the reduction caused by skin stimulation of the splint on the extensor surface of the forearm and hand?

Although splinting and the use of a suspension sling would not be appropriate for all persons with Rett syndrome, the presence of abnormal flexor tone, joint instability, and stereotypic hand behaviors would suggest that such a treatment approach would be indicated. For this student, this approach has opened up a new world, one she is just beginning to explore. ▲

Acknowledgment

I thank Cynthia Schonberg and Barbara Ferrarotti for their support.

References


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