

Hand management in Rett syndrome

Hand related problems in Rett syndrome

Repetitive non-purposeful hand movements are very evident in Rett syndrome and may include one or more of the following actions: mouthing, washing, wringing, clapping, tapping, and/or finger manipulation. Intentional hand movement can be further complicated in the disorder by tremors, uneven muscle tone, spasticity, and apraxia i.e., difficulty in planning and co-ordinating movement.

Hand management initiatives

It is important to realise that, in general, Rett syndrome individuals cannot consciously control their hand movements. Even so, methods of reducing the latter can be utilised to address the needs of the individuals concerned. Those needs may include such things as increasing functional hand use, fostering a greater interaction with the environment, assisting feeding, improving communication capability, and protecting the skin from breakdown and infection.

Splinting is the most common means used to both modify hand movement and promote hand skin care, and should be viewed **as a therapeutic aid and not as a form of restraint**. The type of splints utilised for these purposes include full hand, half hand, or elbow splints. Soft thumb abductor splints can also be used to increase hand function and reduce the possibility of finger deformities.

Other therapeutic aids which have proved effective in the management of hand movement and/or hand skin care include the use of an elastic wrist band or ribbon to hold arm on an armrest, brushing, holding the non-dominant hand, clothing the hands with mittens/socks, verbal commands, wrist weights, distraction by engaging the hands in stimulating activity or where possible, grasping an object.

Resources

Additional information on hand management can be found in the publication 'The Rett Syndrome Handbook' (2007) (2nd edition, pages 121-130) written by Kathy Hunter, and in material retained on this topic by RSAA.

Editor: What follows on this and the following 3 pages is a description of the hand management approach adopted for one Australian Rett syndrome female by her mother, Laraine Barrett, and her daughter's occupational therapist, Dianne Berryman. Dianne also discusses what has appeared in the literature on the use of hand splints in Rett syndrome. For the purpose of confidentiality, the name of the Rett syndrome woman to whom both articles refer, is given as M.

Some long-term adverse effects of stereotypic mouthing/handwringing - One mother's solution

Adverse effects of M's hand activity

My daughter, M, is now thirty years old. Her hands were traumatized by her mouthing which had resulted in calluses, tender soggy skin, and constant fungal infections of the fingertips and fingernails. M's hands were obviously painful as she always resisted attempts to clean or dry them, cut her fingernails, or apply creams and ointments.

Several years ago, elbow splints were introduced on two separate occasions, to help reduce the time she spent mouthing. These splints were put on for 20 minutes at a time, three times a day. They achieved very little, except to make her frustrated and miserable. They had no ongoing or lasting effect and were merely a stop-gap, and for her, an ineffectual one at that.

From the time M was 5 years old, it was noticeable that if her right hand was held at lunchtime or other 'finger food' time, she was then able to feed herself quite capably with her left (dominant) hand. This assistance showed that her gross and fine motor skills could be developed, giving her freedom to choose her food and eat at her own pace.

Her stereotypic mouthing and handwringing resulted in constant dribbling and reduced M's capacity to have functional use of either hand. In addition, the odour caused by the saliva had become extremely offensive and was also a cause for a concern.

There were several other issues arising from her stereotypic mouthing and handwringing:

- chronic fungal infection of the fingers/fingernails;
- saliva on M's clothes adding to the odour;
- hypersensitivity of the fingers/fingernails due to the pain of the fungal infections;
- problems with manicure/maintenance of the fingernails, due to M's refusal to cooperate;
- ongoing soft-tissue damage to the fingers caused by her teeth during her mouthing;
- 4 medical procedures to reduce callus development on her hands/fingers.

Hand management in Rett syndrome

Some long-term adverse effects of stereotypic mouthing/handwringing - One mother's solution

Treatments

Staff caring for M in the community residential unit in which she lives, tried herbal remedies and changes in her diet, to help in eliminating the strong odour caused by the saliva. Nothing had worked. When on a visit to the local general practitioner (GP), staff asked for advice on the saliva odour. The GP referred M to a medical specialist with a view to an opinion on removing some of the salivary glands in her mouth as a means of resolving the issue.

I felt that this was an unnecessarily aggressive and invasive solution. It was a case of treating the symptoms rather than the problem itself. As I saw it, the problems all stemmed from the mouthing and handwringing.

Introduction of a hand splint

Not happy with the recommendation of surgery, early last year I arranged a referral to an occupational therapist (OT) specializing in 'hand therapies'. M and I both attended the appointment. I explained the problem and outlined an idea I had formulated for an individual hand splint for the right hand and forearm.

The OT was extremely enthusiastic about the splint and enlisted a paediatric OT to join in our brainstorming.

The first splint which had two parts, was made of a plastic substance, which was porous to allow the skin to breathe. The material was cut to suit M's right hand/arm size, then heated and softened to enable it to be formed to her hand/arm contours. A lower splint reached from under the tips of the fingers to the forearm, while an upper splint was also fitted to encapsulate the hand/forearm.

Slits were made in the lower splint to allow for a soft padding material to hold M's fingers in a natural posture, separate from each other.

How was the splint applied?

A cotton sleeve was first placed over the wrist and arm. While the lower splint was held in place, M's fingers were positioned in their separate padded and designated positions. The upper splint was then placed over the hand/forearm, after which, padded material, fastened with velcro, was used to enclose both splints. A light sleeve zipper held the whole splint in place.

To prevent the healing process from being jeopardized, the OT designed a soft mitten to be worn on the left hand at night in order to reduce mouthing. At this time, M's hands were soft, red, sore and callused, and the incumbent fungal infection was present.

Care plan

The OT's care plan stated that the splint be used during waking hours on the right hand, and taken off at least three times a day to allow for washing of the hand, treatment of the fungal infection, and massage. As stated earlier, a soft mitten was to be placed on the left hand at night.

Positive outcomes

Two weeks after initiating the plan, M's hands were clearly healing. The tender, sore areas on the fingers were showing clean healthy skin. At 4 weeks, the fungal infection was less evident.

Now, fifteen months after the splint was introduced, the following positive outcomes have been achieved:

- mouthing and handwringing are considerably reduced;
- dribbling is minimal;
- M has demonstrated an improvement in functional use of the left hand;
- the strong odour associated with the saliva has been eliminated;
- the chronic fungal infection of the fingers/fingernails has not recurred;
- hypersensitivity of the fingers is reduced to more normal levels;
- problems with manicure/maintenance of fingernails are minimal;
- soft-tissue damage to the fingers has healed and not recurred;
- no further callus development on the hands/fingers.

Unexpected benefits

M has become more relaxed and is less agitated; she is able to relax her left hand on request; she is able to place her [relaxed] left hand on her knee or on a table when requested; she signs "hands on your knees - for NO" and "hands to your chin - for YES" and both of these communications are now being more clearly understood by family and carers.

Splint modifications

Four weeks after initial use, the OT re-evaluated the splint and it was redesigned to be less bulky.

At present, its design has been refined to be a less restrictive, smaller version, with the upper splint having been eliminated.

Conclusion

The splint that I have described, which was designed specifically to address M's issues, has proven to be a successful and rewarding therapeutic aid for her.

Some long-term adverse effects of stereotypic mouthing/handwringing and one mother's solution

Conclusion (cont.)

M is a happier and more relaxed young woman, and is able to "engage" with family and carers at a higher level than previously. Her ability to communicate has improved, and all those associated with her have commented on the improvement in her general demeanour and well-being.

I am very grateful to Ms D. Berryman, occupational therapist, for her expertise, assistance and enthusiasm, with our [successful] little project.

Laraine Barrett.

Therapist's perspective on hand splints for Rett syndrome: A discussion of the literature

Background

I am an occupational therapist working in the Acute service of Southern Health in Victoria, Australia. Fifteen months ago, I received a referral requesting hand splints for a 30 year old woman with Rett syndrome. For a hand therapist in an acute setting, this was not a common request, and subsequently led to a search of relevant information to determine the likely effectiveness of splints as a therapeutic intervention in this situation.

Rett syndrome is a disorder characterised by sudden loss of motor and cognitive abilities in young girls between 7 and 18 months of age. The most characteristic observable features that differentiate it from other forms of developmental delay, are the stereotypic handwringing, mouthing and biting behaviours, with concurrent loss of previously acquired hand function (2, 3). Hand mouthing and biting can also lead to skin maceration of the fingers and back of the hands (2).

Attempts to control the stereotypic hand movements with behavioural modification techniques or medications, have not been successful (3). However, studies have shown that elbow or hand splints can be beneficial in some cases.

Investigation of the use of hand/wrist splints

My client (M) had previously trialled elbow splints, but her mother reported that these had not been satisfactory to carers. I therefore investigated hand and wrist splints only. Three articles specific to hand/wrist splints were found, though none were later than 1992.

Naganuma and Billingsley (1988) studied the effects of bilateral hand-based thumb abduction splints on three girls aged 13 to 16 years. Their study, conducted over three months, found that stereotypic movements decreased on application of splints, but this result was not carried over when splints were removed. For the one girl who had some limited hand function in self-feeding prior to trial of the splints, an increase of purposeful hand use was observed with application of splints, but not for those girls who had no purposeful hand use prior to the

Investigation of the use of hand/wrist splints

A replication of this study by Tuten and Miedaner (1989) did not find any benefits to application of hand splints in two subjects. However, the girls in this second study were 5 years old and at a different stage of the syndrome (with reference to Hagberg and Witt-Engerstrom's staging system of skill levels in Rett syndrome). It was proposed that this may impact on the success of the intervention.

The third study (Kubas, 1992) was a single case report of a 20 year old woman, assessed over a three year period. In this case, bilateral wrist splints were used to stabilise the wrists in neutral. Thumb abduction components thumb/finger based splints were added to the program later to facilitate functional hand use. Over the three year period, in combination with an overhead deltoid sling and an ongoing therapy program, the client developed increased self-feeding hand skills and maintained the decrease in stereotypic hand movements, even without the splints on.

In these studies, it was proposed that the splints worked by blocking a component of the total stereotypic movement pattern (wrist and thumb positions appeared to be critical components). It was also suggested that the sensory stimulation of the splint may fulfil the need that is being met by the handwringing or mouthing behaviours (Kubas, 1992; Naganuma and Billingsley, 1988).

Some possible factors the literature suggested be considered when assessing the possible benefits of splinting for Rett syndrome, were:

- preferably adolescent or older;
- preferably some functional hand use prior to splinting;
- need to splint for an extended length of time (years);
- need for progressive reassessment of client's needs/goals with subsequent revision of splints to facilitate increased functional use.

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A therapist's perspective on hand splints for Rett syndrome: A discussion of the literature (cont.)

M's stereotypic hand movement issues

When applying these findings to my patient M, the priority issue was the self injurious results of her stereotypic hand movements. Her constant hand mouthing/biting had resulted in chronic welts and skin break-down on her hands and fingers, while the tightness of her handwringing had resulted in laxity of the joints in her fingers making them susceptible to hyperextension and dislocation.

Methods and outcomes

The initial splint trialled was a unilateral bivalve resting splint for her non-dominant hand, supporting M's thumb and fingers in extension, and wrist in neutral, and protecting the dorsum of her hand from mouthing.

It was soon found that the same effect could be achieved with just the volar component of the splint and a light material cover to protect the straps from becoming wet from mouthing. The rationale was to break up the flexor pattern with thumb and wrist extension, and to protect the lax joints of the fingers from further wringing/twisting. It was also hoped that blocking the flexor pattern on one hand would have a carryover effect on the contralateral hand, and that preventing the hand-wringing would free the dominant hand to allow increased use in self-feeding (M previously had some limited use of this hand).

After 1 year of splinting, with some modifications to reduce heat/sweating, and altered strapping for better support of the fingers, there has been a significant observable improvement in M's skin integrity. Her carer also reports that restraining the non-dominant hand has allowed increased functional use of the other hand.

Conclusion

In conclusion, there is some evidence hand splints can be beneficial in assisting to manage the stereotypic hand movements associated with Rett syndrome in some cases. However, it appears that splints need to be applied over an extended length of time to be effective and may not be beneficial in all cases. Splints also need to be part of an overall therapy plan if increased hand skills are to be obtained.

Dianne Berryman.

References:

1. Kubas, E. (1992) - Use of Splints to Develop Hand Skills in a Woman with Rett Syndrome. *The American Journal of Occupational Therapy*, 46 (4). pages 364-368.
2. Naganuma, G. & Billingsley, F.(1988) - Effect of Hand Splints on Stereotypic Hand Behaviours of Three Girls with Rett Syndrome. *Physical Therapy* 68 (5). pages 664-671.
3. Tuten, H. & Miedaner, J. (1989) - Effect of Hand Splints on Stereotypic Behaviour of Girls with Rett Syndrome: A Replication Study. *Physical Therapy* 69 (12). pages 1099-1103.

Rettnet references

Editor: The Rettnet, which is a Rett syndrome discussion database on the internet, is a forum in which the majority of contributors are parents of Rett syndrome children. A brief selection of their comments on hand management follows.

Weights

Elaina, 4 years, seems to prefer that we weigh down her hand. It must be her left one as she gets very annoyed if we immobilise her right. As a result, she is more capable of doing things like eating or switch use. This strategy, which now uses a 6 pound weight, works better for her than splints.

Elbow splints

I am all for arm splints. Before Becky starting using them, she hardly had any hand use at all. Now, when wearing them, she can press buttons on her communication device to ask for food, and indicate her need to go to the toilet. Becky, now 7 years old, hated the arm splints at first, but over time she has gotten used to them. I feel that she actually likes them on at times to give her handwringing a rest.

Hand splints

My 13 year old daughter, Kimberley, is a hand mouther. She wears hand splints, usually during the morning and at meal times. If she happens to be unwell, we always let her have her hands as they comfort her.

Not only do Kimberley's splints cut down the wear and tear on her hands, they enable her to participate in activities such as art. The most critical issue for her has been tightening of her hand and finger muscles which have been developing in recent years. Her hand splints intervene in this process by stretching her hands out in a semi-flat position. I can only imagine how tight her hands would be now if she hadn't been wearing splints since she was 2 years old.

Hot water bottle

We have discovered a very simple way to keep Angie's hands still. We put a hot bottle filled two thirds with very warm water (with the extra air squeezed out), either resting on her lap or on the table in front of her. She loves the feel of it and will let her hands rest on it, relaxed and still, for 15-20 minutes. She likes to squeeze it too.

Hand sores

The healthy hand routine that we use for our daughter is morning and evening anti-bacterial hand washes, together with almost daily soakings in vinegar water followed by the application of vitamin E cream. Prior to adopting this routine, her hands were horribly sore, particularly in winter.