Addressing the Needs of Students with Rett Syndrome

Antonis Katsiyannis

Jennifer S. Ellenburg

Olivia M. Acton

Gregory Torrey

Ana is a nonverbal, ambulatory child who demonstrates a consistent behavior pattern of screaming, hand mouthing/biting, crying, wandering, and posturing (standing with head tilted up towards the ceiling) while standing. She does not demonstrate any independent object manipulation skills and requires continuous physical assistance, prompts, and cues to engage in any activities. Her records indicate that Anna developed normally until 18 months of age, when she began displaying "significant crying spells." After 18 months, she showed a marked deterioration in her development and began experiencing generalized motor seizures. (Ana—not her real name—is a child with Rett syndrome currently served in a midwestern state developmental facility.)

Rett syndrome affects 8,000-10,000 females in the United States. Students with Rett syndrome are normally classified as having severe or profound mental retardation (American Psychiatric Association [APA], 1994). Given current trends in special education, along with the Individuals with Disabilities Education Act Amendments of 1997 (IDEA 97), emphasizing the

What Is Rett Syndrome?

Rett syndrome, evident only in females, is characterized by the "development of multiple specific deficits following a period of normal functioning after birth" (APA, 1994, p. 71). Attempts to identify the cause(s) of Rett syndrome have been largely unsuccessful until recently. Because Rett syndrome is evident only in females, scientists have maintained their focus on genetic causes (Willard & Hendrich, 1999). These efforts have resulted in the isolation of a mutated gene as the cause of this disorder. With the discovery of the gene, researchers can now focus on designing a test for early diagnosis and develop possible treatment options before the birth of these children (Amir et al., 1999).

To be classified as having this disorder, a person must exhibit all the following characteristics:

- Apparently normal prenatal and perinatal development.
- Apparently normal psychomotor development through the first 5 months after birth.
- Normal head circumference at birth.

In addition, the following characteristics must be present following the period of normal development:

- Deceleration of head growth between ages 5 and 48 months.
- Loss of previously acquired purposeful hand skills between ages 5 and 30 months with the subsequent development of stereotyped hand movements.
- Loss of social engagement early in the course.
- Appearance of poorly coordinated gait or trunk movements.
- Severely impaired expressive and receptive language development with severe psychomotor retardation.

Physical and behavioral difficulties, along with delays in normal development, are typical characteristics of people with Rett syndrome. Some of the physical problems they experience include seizures, inability to digest food, loss of purposeful hand movements, and curvature of the spine. Stereotypic behaviors that occur include hyperventilation, facial grimacing, body rocking, teeth grinding, and hand wringing (Smith, Klevstrand, & Lovaas, 1995).

Stereotypic hand movements with loss of functional hand skills, however, are the most distinguishing characteristics of Rett syndrome. Types of hand stereotypes 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 (Sharpe, 1992)

Further, problems with chewing, swallowing, choking, and vomiting complicated by breathing abnormalities, and impaired hand use result in prolonged feeding sessions, gastronomy feeding, and potentially death from malnutrition. Finally, characteristics, such as changes in social contact, withdrawal, and stereotypic movements, typify autistic-like behaviors (Stewart, Brady, Crowe, & Naganuma, 1989).

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expanded use of placements in the least restrictive environment for students with disabilities, public schools will address the needs of people with rare syndromes, such as Rett syndrome, in school-based programs. For instance, in the 1996-97 school year, less than 2% of students with disabilities ages 6 to 21 were educated in residential, hospital, or home-bound environments (U.S. Department of Education, 1999).

An informal search on CARL Uncover (**http://uncweb.carl.org:80/cgi-bin/cw_cgi?28358+setGateway**) revealed that since 1988, more than 235 manuscripts have been published on Rett syndrome—almost exclusively in medical journals. Only a nominal number of articles have appeared in educationally oriented journals (see Table 1 for key resources)

We believe that school personnel, especially teachers, must be informed of behavioral and educational interventions regarding students with Rett syndrome and work closely with physical and occupational therapists so that appropriate programming will be available in the least restrictive environment.

General Interventions

Van Acker (1991) underscored the importance of physical and occupational therapists' role in working with children with Rett. Interventions, although not successful in altering the course of the disorder, have been successful in treating the symptoms to maintain or improve function, prevent deformities, provide correct positioning, and keep the children aware of their environment [see also International Rett Syndrome Association (IRSA) **www.rettsyndrome.org**].

As shown in Table 2, (page 76), selected interventions for symptoms such as apraxia, spasticity, ambulation, scoliosis, seizures, stereotypic behavior, communication, feeding and nutrition, and self-injurious behaviors are provided. The table also provides appropriate, relevant Web pages and specific literature. Further, individualized education program (IEP) goals and objectives must be functional, and adaptive equipment, modifications, communication devices, or a one-on-one aide to achieve stated goals must be included in the IEP, as needed (see Table 3, page 77, for sample IEP objectives).

Specific Interventions

This section provides a review of specific research related to interventions used with children with Rett syndrome. We believe that such a review is essential, given the challenges associated with the syndrome and the need for educational personnel to implement research-validated practices. In passing IDEA '97, the U.S. Congress noted that IDEA has been successful in ensuring that qualified students have access to a free, appropriate public education (FAPE) and in improving their outcomes. As Congress noted, the "Act has been impeded by low expectations, and an insufficient focus on applying replicable research on proven methods of teaching and learning for students with disabilities" [Section 601 (C) (4)]

Communication Abilities

Woodyatt and Ozanne (1992) investigated the communicative intent of the interactive behaviors of six females with Rett syndrome through observation. These researchers found that the nonverbal behaviors exhibited by the children in the study were preintentional. Caregivers, therefore, should recognize nonverbal behaviors as possible attempts to communicate and seek to determine the intent of these behaviors when responding to them (Woodyatt & Ozanne, 1993).

Some children were successful in communicating with head switches.

A 3-year-old child with Rett syndrome was presented with four computer games over the course of 8 months that elicited specific behaviors, including reaching, kicking, visual regard, and vocalization. These games allowed the child to express preference and were intended to improve the child's social-interaction skills. As a result of the child's interactions with the computer games, Watson, Umansky, Marcy, and Repacholi (1996) concluded that the child displayed the ability for preference and intention when provided with these stimuli.

Finally, a preschool-age child with Rett syndrome was presented with two switches that could be activated by her head or hand. After pressing the two switches, the child was presented with toys as part of contingent reinforcement regimen. Data revealed that the child quickly mastered the use of the head switch; however, use of the hand switch was less frequent and more variable (Sullivan, Laverick, & Lewis, 1995).

Stereotypic Movements

Wehmeyer, Bourland, and Ingram (1993) investigated environmental consequences that maintained stereotyped behaviors of two children with Rett syndrome. Participants were observed in a random sequence, and the investigators recorded the frequency of hand behaviors. Four different conditions were tested: alone (no demands or activities were presented), demand (received continual verbal and physical prompting to engage in an activity), attention (physi-

Table 1. Selected Resources on Rett Syndrome

Comprehensive Web pages—characteristics, therapeutic, and educational interventions, links…	International Rett Syndrome Association www.rettsyndrome.org
Comprehensive reviews of Rett-related literature	Perry (1991) Van Acker (1991)
Review of interventions	Hanks (1986, 1990)

Table 2. Selected Interventions and Resources for Rett Syndrome Related Symptoms

Symptom	*Interventions*	*Resources*
Apraxia-Ataxia	Use of a therapy ball Balance stimulating floor activities Rotation and weight shift activities	Van Acker (1991) Hanks (1986) www.cec.sped.org/faq/apraxia.htm
Spasticity	Hydrotherapy (e.g., movement in the water, range of motion) Tone reduction activities (e.g., rotation, weight shift)	Hanks (1986)
Ambulation	Weight-bearing exercises Walking and stair climbing	Hanks (1986)
Scoliosis	Continuous exercising of muscles often avoided Good positioning should be stressed	Hanks (1986) www.coolware.com/health./medical_reporter/scoliosis.html http://members.xoom.com/hydranenceph/spiral.htm
Seizure	Management through medication (Tegretol)	http://dir.yahoo.com/Health/Diseases_and_Conditions/Epilepsy
Stereotyped hand movements	Splinting Music therapy Highly motivating activities (e.g., toys with bright colors and sound may be helpful to get child to use hands)	Hanks (1986)
Communication programming	Use of communication systems (e.g., eye pointing, communication boards, facial expressions, gestures, and activation of switches)	www.rettsyndrome.org
Feeding and nutrition	Behavioral interventions for finger feeding and modified utensil use Nutritional interventions Proper positioning and tone reduction activities (spasticity) for reducing choking risk	Hanks (1986) www.new-vis.com www.thickandeasy.com
Irritability, self-injurious, aggressive behaviors	Behavioral interventions Calming activities (music, warm baths...) Management through medication for sleeping disturbances (Tranzene)	Van Acker (1991)

cal contact with no access to tasks), and leisure (with access to items used in the demand condition). For the first participant, the behavior was most likely to occur in the demand phase, while the second participant exhibited the behaviors during the alone condition. The attention and leisure conditions were effective in decreasing the occurrence of the behavior.

Self-Injurious Behaviors

Oliver, Murphy, Crayton, and Corbett (1993) examined the effects of four different experimental conditions on the self-injurious behavior (SIB) of a 3½-year-old child with Rett Syndrome. In the first condition, the child was exposed to continuous attention. During this condition, an adult gave the child his or her full attention and attempted to maintain a pleasant atmosphere while playing games, singing, and talking, in addition to other activities. The child also underwent a no-stimulation condition during which time the child was alone in a quiet room. Third, a stimulation condition was used in which the child was given some of her favorite toys and did not receive close adult supervision. Finally, the demand condition consisted of an adult requir-

Table 3. Sample IEP Objectives (adapted from IRSA Web site—www.rettsyndrome.org)

Communication	During morning circle, Julie will use three switches with icons and labels to communicate her choice of songs,independently, on 4 out of 5 trials.
Drinking	When presented with something she likes to drink, Julie will sip liquid through a plastic straw with consecutive swallows, independently, without biting straw, on 3 out of 4 attempts.
Eating	When presented with finger food, Julie will bring finger food to mouth and release finger food into mouth without dropping on 3 out of 5 attempts.
Hand use	During choice making activity, Julie will reach out to touch object or picture as directed by the teacher, independently, on 3 out of 5 attempts.
Mobility	Julie will maintain sitting balance on chair with back, without supports, for 5 consecutive minutes.
Sensory/ Attention to task	During tooth brushing, Julie will decrease number of times she bites on toothbrush to less than 3 per instance over the next 3 weeks. During nap time, Julie will lie quietly on mat for at least 15 minutes over 3 weeks.
Social skills	During morning circle, Julie will touch a friend on the arm to say "hello," independently, at least once over a 3-week period.

Note: Adapted from IRSA Web site—**www.rettsyndrome.org**

ing the child to perform three 5-minute tasks during the session. In four out of nine sessions, the continuous-attention condition resulted in extreme SIB and distress causing the experimenters to terminate the condition. A functional analysis was performed; the investigators deemed the self-injurious behaviors to be a result of reinforcement by sensory stimulation and escape from social interactions.

Similarly, Iwata, Pace, Willis, Gamache, and Hyman (1986) studied two children with Rett syndrome who displayed SIB. These children were subjected to several interventions that were paired with different types of reinforcements. Some of the interventions included differential reinforcement, response interruption techniques, guided compliance, ignoring, and the use of mitts. The most successful intervention for both of the children was differential reinforcement (reinforcement in the presence of one stimulus but not others) paired with response interruption.

Paisey, Whitney, and Wainczak (1993) concluded that a combination of graduated guidance and social and edible reinforcers successfully established stable rates of functional hand movements to activate toys, gross motor responses to verbal prompts, palm grasp and release, and some vocal imitation. Contingent response interruption virtually eliminated hand mouthing during instructional sessions. Differential reinforcement plus response interruption appeared to be superior to hand splints in reducing hand stereotypies.

Stereotypic Behaviors

Sharpe (1992) and Aron (1990) found that stereotypic hand movements were reduced and toy contact increased with the use of the elbow orthosis in subjects with Rett syndrome. In contrast, bilateral hand splints had no obvious treatment effect (Naganuma & Billingsley, 1988). The elbow orthosis, nonetheless, is easy to fabricate, inexpensive, and simple to apply. There are limitations to its effectiveness, however. 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 (Sharpe, 1992). Decreasing the repetitive hand movements can lead to greater alertness and better focus, as well as helping to decrease agitation and self-injurious behavior (IRSA).

Feeding Behaviors

Stewart et al. (1989) concluded that parental concerns involved primarily

Certain interventions have been successful in treating the symptoms to maintain or improve function, prevent deformities, provide correct positioning, and keep the children aware of their environment.

IEPs must include, as needed, functional goals and objectives, adaptive equipment, modifications, communication devices, or a one-on-one aide to achieve stated goals.

feeding-related problems. Therapists improve feeding skills by addressing poor positioning, poor oral-motor control, and impaired functional hand use through evaluation and treatment. Overall, therapists need to be aware of the distinct stage of the disorder to provide the best treatment.

Final Thoughts

Addressing the needs of students with Rett syndrome is an extremely challenging endeavor. Teachers need to be aware of a variety of behavioral interventions found in the literature, focus on individualized approaches, consider contextual factors, work closely with physical and occupational therapists, and employ the least intrusive interventions.

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Antonis Katsiyannis *(CEC Chapter #997), Professor, Special Education, Clemson University, , SC.* **Jennifer S. Ellenburg** *(CEC Chapter #147), Special Education Teacher; and* **Olivia M. Acton,** *Special Education Teacher, Charlotte-Mecklenburg Public Schools, Charlotte, NC.* **Gregory K. Torrey,** *Psychologist Associate, Beatrice State Developmental Center, Beatrice, NE.*

Address correspondence to Antonis Katsiyannis, Clemson University, 407C Tillman Hall, Clemson, SC 29634. E-mail: antonis@clemson.edu

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