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# Communication Skills in Girls With Rett Syndrome

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## Abstract

Rett Syndrome (RS) is an X-linked, neurodevelopmental disorder that occurs primarily in females and causes significant impairment in cognition, motor control, and communication. Teachers and speech-language pathologists (SLPs) encounter girls with RS with increasing frequency as awareness of the disorder increases, yet the literature on clinical interventions with this population is limited. Parents, teachers, and SLPs were surveyed regarding the communication abilities of these girls. Respondents reported that girls with RS use multiple modalities to communicate, with eye gaze the most frequently used modality, followed by picture/symbol systems and body movements. This study provides clinical information for professionals working with girls with RS, and supports the need for additional research on strategies to improve communication in RS.

## Keywords

Rett Syndrome, communication modalities, survey, treatment

Rett Syndrome (RS) is an X-linked dominant neurodevelopmental disorder that affects approximately 1 in 10,000 females (Amir & Zoghbi, 2000). Hallmarks of the disorder are significant communication impairment, severe to profound intellectual disabilities, and poor motor skills (Gillberg, 1997). More than 85% of cases of RS have an identified mutation in the MECP2 gene (Cass, Reilly, Owen, & Wisbeach, 2003). The role of MECP2 in brain development is not fully understood at this time; however, it is hypothesized that an error in the MECP2 gene alters the normal developmental expression of various other genes (Amir & Zoghbi, 2000). The clinical phenotype of girls with RS is changing as a result of advances in genetic testing and greater awareness of the syndrome (Weaving, Ellaway, Gecz, & Christodoulou, 2005). This includes girls with considerably higher cognitive abilities than previously reported. Researchers now describe girls with RS who are capable of intentional communication and verbal language skills (Hetzroni & Rubin, 2006; Skotko, Koppenhaver, & Erickson, 2004; Zappella et al., 2003).

## Description of RS

RS has been described as a staged disorder (Kerr & Engerstrom, 2001) in which the symptoms of the disorder become more evident and change with age. Although there is some dispute that all girls with RS progress through each

stage, the stage model is helpful in describing a general progression of the disorder. In the initial stage (birth to approximately 1 year), many girls are reported to have a normal year of life during which they begin to acquire early speech and walking skills (Cass et al., 2003). Sometime after the child's first birthday the symptoms of RS become evident as the girl moves into Stage 2 of the disorder. In this stage, referred to as the regression stage, previously acquired skills are lost, particularly motor and language skills, and seizures may begin. Respiratory, sleeping, and feeding problems also may emerge (Naidu et al., 2003). In Stage 2, girls may appear socially withdrawn and receive the diagnosis of autism. Repetitive, nonfunctional hand movements begin. These stereotypic hand motions affect the child's ability to interact with and learn from her environment. Apraxia, a motor disorder of voluntary movement, becomes evident in Stage 2. This difficulty can affect all planned motor movements, including limb movements and speech (Hetzroni, Rubin, &

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Konkol, 2002), making it especially challenging for teachers and therapists to adapt educational materials and provide access to technology. Initiation of voluntary movement may be delayed, often as long as 30 s (Bartolotta, 2005). There is evidence that accurate responses are produced by girls with RS when their communication partners provide sufficient wait time after asking a question (Koppenhaver, Erickson, Harris, et al., 2001; Koppenhaver, Erickson, & Skotko, 2001; Skotko et al., 2004). This second stage of RS may last for 1 or more years, typically ending between ages 5 and 10 years (Cass et al., 2003).

In Stage 3, referred to as the pseudostationary phase, there is typically an improvement in symptoms (Cass et al., 2003). Agitation and problems in sleeping may diminish, and the symptoms typical of autism (i.e., poor eye contact, social withdrawal) also seem to improve. There may be an improvement in communication and motor skills during this time (Jacobsen, Viken, & Von Tetzchner, 2001). A girl with RS may remain in this third stage well into adulthood. The fourth stage of the disorder, sometimes referred to as the nonambulant phase, begins around the time of puberty. This stage is characterized by an increasing rigidity throughout the body, with a loss of walking skills for some girls and a decrease in repetitive hand movements (Cass et al., 2003). Eye contact often improves during this stage. If a girl with RS is relatively healthy, she may live well into adulthood (Jacobsen et al., 2001), although sudden death of unknown origin, particularly during sleep, is reported in some girls with RS. Many girls with RS have accompanying medical conditions that affect their long-term health such as pneumonia, dysphagia, and seizures (Budden, 1986; Budden, Meek, & Henighan, 1990; Cass et al., 2003).

There is little published research on the communication abilities of girls with RS. A retrospective case study of 87 girls with RS confirmed previous findings that girls with RS are profoundly impaired in cognitive and communication skills (Cass et al., 2003). Specific communication strategies used by girls were not described in this study.

When specific adaptations are made, girls with RS may display communicative behavior that is intentional. In a study of augmentative communication training, three girls with RS learned to touch a computer screen to make requests for desired food items (Van Acker & Grant, 1995). The girls displayed a combination of behaviors (i.e., eye gaze, vocalizations, facial expressions, gestures, walking to a desired object or activity) that were reported to be communicative in nature. This observation has been noted in other studies as well, suggesting that unique or "non-standard" behaviors produced by girls with RS may be meaningful and valued as intentional communication attempts (Hetzroni & Rubin, 2006; Sigafos et al., 2000; Skotko et al., 2004). In a study examining meaningful use of eye gaze, three girls with RS were trained to look at a named object when picture communication symbols were

presented on a computer screen. At the end of the training, the girls started to use the symbols during classroom instruction. Though the authors cautioned that their data could not be generalized beyond the observations made in the participants' classrooms, they recommended that additional research be conducted to determine how to effectively train girls with RS to use symbols for communication (Hetzroni et al., 2002).

Skotko et al. (2004) noted that girls with RS learned to communicate in meaningful ways during the context of storybook reading with their mothers. In this study, the researchers provided training to the mothers of four girls with RS to enhance the quality of story reading interactions. The training strategies included attributing meaning to the girls' attempts to communicate, asking communicatively relevant questions, and waiting between 8 and 30 s to allow their daughters to respond. These adaptations resulted in increased numbers of communicative attempts by the girls with RS.

Although there is some evidence that girls with RS can communicate intentionally, a need exists for clinical research that describes the varied communication abilities of this population. Researchers indicate that familiar communication partners have learned to interpret the behaviors of girls with RS as meaningful. The role of the communication partner is key to the evolution of intentionality in the person with a severe disability (Rowland, 2003). In an exchange with a person who has severe disabilities, the communication partner must use inferences and intuition to ascribe meaning to behaviors (Grove, Bunning, Porter, & Olsson, 1999). In the Skotko et al. study (2004), the mothers were asked to assume that their daughters were competent communicators at the start of the study. As part of the training, the mothers were instructed to attribute meaning to their daughters' vocalizations, eye gaze patterns, and gestures. The researchers concluded that by attributing meaning to the girls' initial behaviors, the girls were able to learn to communicate intentionally through a variety of means, including novel methods, by the end of the project.

This study was undertaken in an attempt to understand the perceptions of people who regularly communicate with girls with RS to obtain information about the type and effectiveness of each girl's communication ability. The intention was to build an understanding of the range of behaviors produced by girls with RS as the clinical presentation of the syndrome is changing and more girls with a greater range of ability and skill level are receiving the diagnosis.

## Method

### Participants

Participants were recruited for the study by email posted on the Rettnet, an electronic listserv administered by the

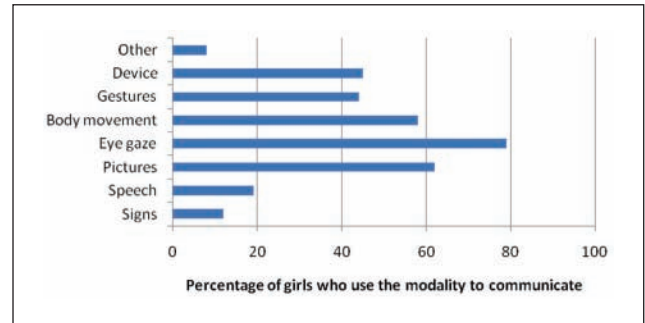
International Rett Syndrome Association (IRSA) and by distribution of fliers describing the study at the annual conference of the IRSA. Parents are most familiar with their daughters' skills and the unique communication behaviors displayed by each girl. Speech-language pathologists (SLPs) and teachers who work with girls with RS become expert in identifying skills and competencies in individual girls in their classrooms. Therefore, a convenience sample of parents of girls with RS and teachers and SLPs who work with a girl with RS served as respondents. A total of 152 persons attempted to complete the survey. Of the surveys, 11 were incomplete at the time the survey was closed so those responses were discarded. A total of 141 completed surveys were available for analysis. Of these, 116 were completed by parents, and 25 were completed by professionals (teachers and SLPs) working with girls with RS. The majority of the respondents (84%) resided in the United States. The other respondents were from eight other countries including Canada, Australia, and several within Western Europe.

The ages of the girls with RS were distributed across the life span. The largest group of girls was within the 4 to 7 year age group (33%), with the remaining girls spread across the other four age groups: 0 to 3 years (15%), 8 to 13 years (24%), 13 to 20 years (17%), and 21 years and older (11%).

## Survey

Validity studies were conducted on a preliminary version of the survey using a panel of seven experts in the field of RS who assessed the content and face validity of the preliminary survey and made suggestions to improve wording and content of the survey. The survey contained three sections. In the first section respondents answered 13 demographic questions regarding themselves and the girl with RS.

The second section of the survey contained 20 statements to measure the respondents' perceptions of the girls' communication skills. Respondents were asked to reply using a 5-point scale (i.e., 5 = *strongly agree*, 4 = *agree*, 3 = *undecided or unsure*, 2 = *disagree*, 1 = *strongly disagree*). The 20 statements were subdivided into four groups, based on the content of the statement. This allowed the researcher to determine the level of agreement or disagreement for each statement and to construct impressions about the respondents' overall responses to similar types of statements. The four groups of statements are: (a) perceptions of expressive communication skills; (b) perceptions of comprehension, cognition, and apraxia; (c) perceptions of communicative effectiveness; and (d) other types of statements. Some of the items queried the availability of materials about RS, participant knowledge of RS, and the effectiveness of therapy. The third section provided the respondents with an open-ended opportunity to share any additional information about the girl with RS and her ability to communicate.



**Figure 1.** Communication modalities used by girls with Rett Syndrome

## Procedures

Volunteers logged onto a URL that was housed on a secure server at Seton Hall University to complete the survey. All responses were anonymous and confidential. Data collection occurred over a 2-month period. The survey was available only in English.

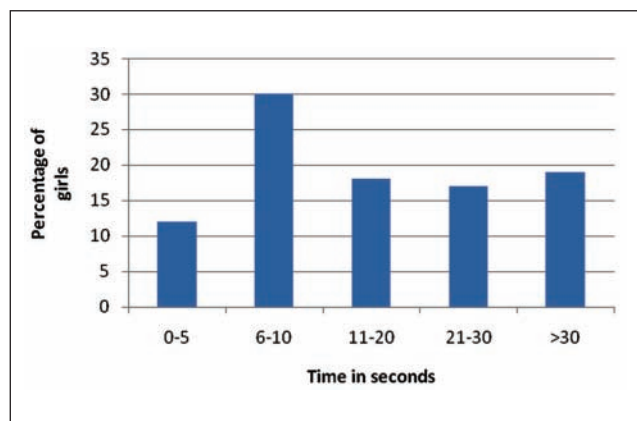
Data obtained from each of the 141 complete surveys were exported from the web server directly to the Statistical Package for Social Sciences (SPSS 12.0) for analysis. The data were examined to determine which statistical methods would be most appropriate for describing the demographic information and for analyzing the relationships between the independent variables (answers to demographic questions) and the dependent variables (answers to Likert-type scale questions). Data analysis included both descriptive and nonparametric statistics. In the analysis of the responses to the Likert-type scale questions, percentage, standard deviation, mean, and mode are all reported to describe the central tendency and variability in the data.

## Results

### Communication Modality Use

Figure 1 contains the eight modality choices given in the survey and the percentage of girls who were reported to use each. Respondents reported each girl used an average of three different modalities to communicate information to others. Eye gaze in some form was reported to be the most frequently used communication modality, as it was used by 78.9% of the girls with RS. Body movement and pictures or symbol boards were the next most frequently reported communication modality, used by 56.3% and 61.3% of the girls with RS, respectively. Examples of "other" behaviors produced by girls with RS that are potentially communicative included crying, hyperventilating, and laughing (Hetzroni & Rubin, 2006).

Of those girls who were able to access a communication device, 60.3% used touch with hands or fingers and 57.4%



**Figure 2.** Length of time required to produce a response

used some type of eye gaze. A head pointer was reported to be used by 2.8% of girls, and other types of access (not specified) were used by 5.0% of the girls. Of the entire sample, 11.3% of the girls with RS reportedly did not use any type of communication system.

### Response Time

Respondents were asked to report how long it took for a girl to generate a response to a stimulus (Figure 2). Of the entire group of girls, 57% required a delay of 11 s or more to generate a response.

### Therapy History

More than 90% of the girls with RS were reported to have received speech, physical, and occupational therapies, and 50% had received an augmentative communication evaluation. Just more than one third (38%) received music therapy. A total of three girls (2.1%) have not received any of these therapeutic services. Two of these girls resided in the United States and one resided in Portugal.

### Educational Programming

More than 84% of the girls were currently enrolled in some type of educational program. At least 61% were in a special education classroom, 18% were included in general education classrooms with typical peers, and 5% were in an adult day program. The remainder ( $n = 11$ ), the majority of whom were adults, were not in any type of program at the time of the survey.

### History of Speech Use

Of the girls, 70% were reported to have used oral speech at some time in their lives. Of these girls, 86% experienced a

**Table 1.** Responses to Statements Regarding Perceptions of Expressive Communication Skills

| Statement           | <i>M</i> | <i>SD</i> | <i>Mdn</i> | Mode |
|---------------------|----------|-----------|------------|------|
| Communicates using: |          |           |            |      |
| Spoken single words | 2.09     | 1.43      | 1          | 1    |
| Spoken sentences    | 1.41     | 0.90      | 1          | 1    |
| Vocalizations       | 4.02     | 1.30      | 4          | 5    |
| Sign(s)             | 1.87     | 1.15      | 1          | 1    |
| Pictures or symbols | 3.65     | 1.40      | 4          | 4    |
| Gestures            | 3.12     | 1.50      | 4          | 4    |
| Eye gaze            | 4.33     | 0.99      | 5          | 5    |
| Body movement       | 3.79     | 1.34      | 4          | 5    |

loss or regression in speech skills. For 59%, this regression occurred between 1 and 2 years of age. Approximately 11% were younger than 1 year of age at the time of regression, 20% were between ages 2 and 3 years, and 10% were older than 3 years of age. Of the girls who lost speech, 44% had fewer than 5 single words before the regression occurred, 37% had acquired between 10 and 50 single words, 11% used short phrases, and 6% had developed sentence use before the regression. Of the 6 girls who were reported to have acquired sentence use before the regression, 3 of them were older than 3 years when the loss of speech occurred. The remaining three girls were between 1 and 2 years ( $n = 1$ ) and between 2 and 3 years ( $n = 2$ ) of age at the time of speech loss. Of those who were reported to have a history of speech use, 46% are reported to speak at the current time, most using 10 single words or fewer. The words that were reported to be used were typically names of family members (e.g., *Mom*, *Dad*) and other single words that can be used in multiple contexts, such as *no*, *yes*, *more*, *bye*, *go*.

### Analysis of Likert-Type Scale Statements

Table 1 presents the distribution of the data for the perceptions of the expressive communication skills statements. There were three questions in this section, for which the mode was 5 (*strongly agree*). Of respondents, 57% chose *strongly agree* as a response to the eye gaze question, 46% chose *strongly agree* for the vocalization question, and 37% chose *strongly agree* for body movement. In summary, the respondents perceived that most girls with RS communicate with eye gaze, vocalizations, and body movements and far fewer girls use spoken words or sentences and signs.

The data for the perceptions of comprehension, cognition, and apraxia statements are presented in Table 2. There were two questions in this section, for which the mode was 5 (*strongly agree*). Of the respondents, 73% strongly agreed with the statement that girls with RS understand at least 10 spoken words. Strong agreement with the statement that apraxia limits the ability to communicate was indicated by



**Table 2.** Responses to Statements Regarding Comprehension, Comprehension, and Apraxia

| Statement                                 | M    | SD   | Mdn | Mode |
|-------------------------------------------|------|------|-----|------|
| Can read 1 or more single words           | 2.66 | 1.52 | 3   | 1    |
| Understands at least 10 spoken words      | 4.60 | 0.76 | 5   | 5    |
| Follows one-step commands                 | 3.79 | 1.18 | 4   | 4    |
| Has a significant cognitive impairment    | 2.93 | 1.28 | 3   | 4    |
| Apraxia limits her ability to communicate | 4.35 | 0.91 | 5   | 5    |

**Table 3.** Responses to Statements Regarding Perceptions of Communication Effectiveness

| Statement                                                | M    | SD   | Mdn | Mode |
|----------------------------------------------------------|------|------|-----|------|
| Familiar people can interpret her communication          | 4.41 | 0.79 | 5   | 5    |
| Unfamiliar people cannot interpret her communication     | 3.77 | 1.03 | 4   | 4    |
| Most people accurately estimate how much she understands | 2.17 | 1.08 | 2   | 2    |
| Most people underestimate the abilities of girls with RS | 4.30 | 1.19 | 5   | 5    |

58% of the respondents. Of note is that the standard deviation for each of these questions was fairly small ( $< 1$ ), indicating little variability in responses of respondents to these questions. In summary, interpretation of the data presented in Table 2 indicates that respondents perceived that most girls with RS understand at least 10 words and that apraxia is a significant barrier to communication.

The data for the perceptions of communication effectiveness statements are presented in Table 3. There were two questions in this group, for which the mode was 5 (*strongly agree*). Of respondents, 55% strongly agreed with the statement that familiar people can interpret the communication of a girl with RS. Of respondents, 65% strongly agreed with the statement that most people underestimate the abilities of girls with RS. In summary, from the data presented in Table 3 it can be surmised that the respondents perceived that familiar people are the ones who are most likely to accurately interpret the communication of girls with RS and that unfamiliar people have difficulty interpreting the communication behaviors of girls with RS. Respondents also perceived that most people do not accurately estimate how much a girl with RS can understand and will underestimate the abilities of girls with RS.

The distribution of the responses to the remaining three statements is presented in Table 4. Of respondents, 44% strongly agreed with the statement that they had confidence in their skills in working with girls with RS. Of respondents, 47% agreed with the statement that there is information available to help them work with girls with

**Table 4.** Responses to Statements Regarding Perceptions of Effectiveness of Therapy for RS, Respondent Skill Level in Working With RS, and Availability of Materials on RS

| Statement                                          | M    | SD   | Mdn | Mode |
|----------------------------------------------------|------|------|-----|------|
| Communication skills have improved with therapy    | 4.0  | 0.95 | 4   | 4    |
| Confidence in skills in working with girls with RS | 4.19 | 0.91 | 4   | 5    |
| Availability of information on working with RS     | 3.84 | 1.07 | 4   | 4    |

RS, and 41% agreed that the communication skills in girls with RS have improved as a result of therapy. In summary, the data presented in Table 4 can be interpreted to indicate that respondents perceive that their skills in working with girls with RS are strong, that there is a great deal of information available publicly about RS, and that therapy is helpful for improving the communication skills of girls with RS.

Analyses were conducted using nonparametric statistics to determine if there were significant differences between the responses of the parent group and professional group to the 20 perceptions of communication statements. The  $\chi^2$  test of homogeneity was utilized to identify relationships between responses using cross tabulations. Kendall's tau-b also was calculated to determine the strength and direction of each association. Significant differences ( $p \leq .05$ ) between the two groups were identified for the five statements in Table 5. There was no significant difference between perceptions of parents and professionals for the remaining 15 statements. For all of these associations, the results must be interpreted with caution, however, as there were numerous cells that had fewer than five responses per cell.

In summary, parents were significantly more likely than professionals to agree that a girl with RS used single words and gestures to communicate and that the girl understood at least 10 spoken words. The relationship between role of respondent and the response to the statement was not particularly strong according to the Kendall's tau-b analysis, suggesting that other factors may play a role in the respondent's choice of answer. A strong relationship was evident in the responses to the statement "familiar people can interpret her communication attempts" and the role of the respondent. The Kendall's tau-b value was significant and suggested a stronger association between the variables than for the other responses. Specifically, parents were significantly more likely than professionals to agree or strongly agree that familiar people can interpret the communication attempts of girls with RS. A relatively strong relationship also was evident between the role of the respondent and the response to the statement "most people underestimate

**Table 5.** Significant Differences Between Perceptions of Parents and Professionals in Response to Communication Perception Statements

| Communication perception                                 | Pearson $\chi^2$ |    |                          | Kendall's tau-b |             |
|----------------------------------------------------------|------------------|----|--------------------------|-----------------|-------------|
|                                                          | Value            | df | Asymp. sig. (two-tailed) | Value           | Approx sig. |
| She communicates using spoken single words               | 10.071           | 4  | .039*                    | -.052           | .437        |
| She uses gestures to communicate                         | 13.028           | 4  | .011*                    | -.088           | .183        |
| She understands at least 10 spoken words                 | 8.080            | 3  | .044*                    | -.147           | .094        |
| Familiar people can interpret her communication attempts | 23.882           | 4  | < .001*                  | -.351           | < .001*     |
| Most people underestimate the abilities of girls with RS | 16.720           | 4  | .002*                    | -.268           | .003*       |

\* $p < .05$ .

the abilities of girls with RS.” The Kendall’s tau-b value was significant, which suggested a strong negative association between the variables. Here again parents were more likely than professionals to strongly agree that most people underestimate the abilities of girls with RS.

To further explore the perceptions of the respondents, additional associations between responses to the perceptions were studied. A significant association was demonstrated between history of speech use (Question 10 in Section 1 of the survey) and the response to the statement “she communicates using spoken single words,”  $\chi^2(8, N = 139) = 24.465$ ,  $p = .002$ . A significant and strong negative relationship was identified between the variables  $\tau = -.336$ ,  $p = .000$ . This indicated that a girl who achieved a level of oral speech use at some time in her history was more likely to use single words at the time the survey was completed.

The relationships between use of spoken single words along with other communication modalities were explored. Strong, positive, and significant relationships were found between responses to “she communicates using spoken single words” and 7 of the 19 other statements (Table 6). This suggests that respondents who agreed that the girl with RS used single words also were likely to agree that she uses multiple modalities to communicate, and that includes gestures, signs, and pictures, symbols, or objects. The girl with RS who uses single words also is likely to use at least a few sentences to communicate and can follow one-step commands. As expected, a significant and strong relationship

**Table 6.** Significant Relationships Between Responses to “She Communicates Using Spoken Single Words” and Responses to Other Communication Perception Questions

| Communication perception                                     | Pearson $\chi^2$ |    |                          | Kendall's tau-b |             |
|--------------------------------------------------------------|------------------|----|--------------------------|-----------------|-------------|
|                                                              | Value            | df | Asymp. sig. (two-tailed) | Value           | Approx sig. |
| She communicates using spoken sentences                      | 109.491          | 16 | < .001*                  | .592            | < .001*     |
| She communicates using sign(s)                               | 48.482           | 16 | < .001*                  | .334            | < .001*     |
| She uses pictures or symbols or other objects to communicate | 33.639           | 16 | .006*                    | .282            | < .001*     |
| She uses gestures to communicate                             | 55.700           | 16 | < .001*                  | .324            | < .001*     |
| She use body movement to communicate                         | 26.791           | 16 | .044*                    | .259            | < .001*     |
| She follows one-step commands                                | 26.713           | 16 | .045*                    | .253            | < .001*     |
| She has a significant (severe) cognitive impairment          | 32.504           | 16 | .009*                    | -.147           | .047*       |

\* $p < .05$ .

was identified between reported single word use and a significant cognitive impairment. This suggests that respondents who were more likely to agree that the girl with RS uses single words were less likely to agree that she had a significant cognitive impairment.

In the text responses that were given in Section 3 of the survey, respondents were noted to describe apraxia as a factor in limiting a girl’s ability to communicate and in increasing the time required for her to produce a response. Therefore, the association between apraxia (motor planning abilities) and cognition and the perceptions of others was explored by analyzing three sets of related variables. All three of these associations were found to be significant (Table 7). Respondents who agreed with the statement that apraxia interfered with communication were significantly less likely to agree that the girl with RS had a significant cognitive impairment. Respondents who agreed with the statement that apraxia interfered with communication were significantly less likely to agree that people were accurately able to estimate how much a girl with RS can understand. Respondents who agreed that there is a relationship between the influence of apraxia on communication, were significantly likely to agree that other people underestimate the abilities of girls with RS. The relationship between this last set of responses was found to be relatively strong,  $\tau = .274$ ,  $p = .000$ , compared to the previous responses.

**Table 7.** Significant Relationships Between Responses to “Apraxia (Motor Planning Problem) Limits Her Ability to Communicate With Others” and Three Communication Perception Questions

| Communication perception                                           | Pearson $\chi^2$ |    |                          | Kendall's tau-b |              |
|--------------------------------------------------------------------|------------------|----|--------------------------|-----------------|--------------|
|                                                                    | Value            | df | Asymp. sig. (two-tailed) | Value           | Approx. sig. |
| She has a significant (severe) cognitive impairment                | 31.658           | 16 | .011*                    | -.125           | .089         |
| Most people accurately estimate how much she knows and understands | 29.811           | 16 | .019*                    | -.108           | .172         |
| Most people underestimate the abilities of girls with RS           | 39.993           | 16 | .001*                    | .274            | < .001*      |

\* $p < .05$ .

The Mann–Whitney  $U$  test was used to determine if significant differences existed between the responses from the parents and professionals. Mean ranks and summed ranks were calculated on the responses to the 20 perception statements. The differences in mean rank between the groups were significant for only 3 of the 20 statements ( $p \leq .05$ ). Parents were significantly more likely than professionals to indicate agreement with the following statements: “a girl with RS uses vocalizations to communicate,” “familiar people can interpret her communication attempts,” and “most people underestimate the abilities of girls with RS.”

### Additional Responses

A total of 91 respondents (65%) provided responses when asked to share any additional information regarding communication skills, such as communication strategies, words or phrases used, any augmentative systems, or anything unique or special about her communication skills. In these additional responses, participants described successful use of multiple modalities for communication. For example, one parent reported, “She communicates mostly by body language, some vocalizations and with her eyes.” A therapist reported on the successful use of multiple augmentative and alternative communication strategies for one girl with RS by writing the following: “She uses an eye gaze board with up to four pictures, and looks at the picture to make a choice, and then at me to confirm. She also uses a Big Mack (switch) to participate in circle time activities.” Another therapist reported,

This young lady with RS is using a Mercury communication system . . . with direct touch access. There are some continuing issues related to some perseverative tapping around the desired item, but has made HUGE gains. She is very motivated by social interactions. Overall level of undesired behaviors has been reduced significantly!

In addition, one parent expressed frustration at those who underestimated her daughter’s responses, with this sentence: “Our girl has so much to say if we only listen with our senses.” Another described her daughter’s inconsistent behavior by stating, “On a good day, or when she is highly motivated, she can respond within 5 seconds. On a more difficult day of time of day, it may take 30 seconds to get the same response.”

### Discussion

The purpose of this study was to expand the body of literature on communication skills in girls with RS. The findings provide preliminary evidence that girls with RS are perceived by familiar communication partners as capable of intentional communication. This is consistent with previous reports of case studies or small group designs (Hetzroni & Rubin, 2006; Skotko et al., 2004). Girls with RS are reported to use multiple modalities to communicate, including eye gaze, body movements, and augmentative communication systems that contain pictures or symbols. Respondents perceived that apraxia, a motor planning impairment, strongly limits the ability of a girl with RS to communicate with others. Apraxia also is perceived to influence consistency and speed of motor response and to influence how well others can judge the cognitive and comprehension skills of a girl with RS. The results of this study are consistent with previous research that has demonstrated that assessing intentional communication is extremely complex in individuals with severe disabilities (Sigafoos et al., 2000). The role of the communication partner in interactions must be considered when designing assessment tools to evaluate intentional behavior in girls with RS.

Eye gaze was reported to be the most commonly used communication modality in this study. Respondents indicated that girls with RS use eye gaze in many different ways. For example, a girl may look intensely at a desired object without establishing mutual eye gaze with her partner. In this circumstance, the burden of communication is on the partner, who must recognize that the eye gaze pattern is intentional and that the girl with RS is using eye gaze to make a request. This behavior should be verified over multiple trials and with different partners. Those persons who interact with the girl with RS could then be trained to identify those instances in which the girl is using her eye gaze



intentionally. This communication modality can be further developed to see if eye blinks (for a yes–no response) can be trained or if a girl can be trained to use mutual eye gaze behavior to first look at a desired object and then look at the partner to acknowledge the request. We have much to learn about mutual eye gaze behavior in RS. There is some evidence that girls with RS are able to use their eyes to point to desired objects but do not use more advanced gaze switching (looking at an object, then gazing at a communication partner) for communicative purposes (Cass et al., 2003). This is an area in need of further clinical exploration for assessment and intervention.

There are a wide variety of eye gaze systems that can be used for augmentative communication. There are several low-tech options that use direct eye gaze at one to two objects or pictures of objects. If appropriate, more complex, nonelectronic eye gaze systems can be established that use one- or two-step processes, depending on the field of choices that are available. Some girls with RS may be candidates for electronic eye gaze systems that can use digital technology to recognize eye movements of the user. It is important that clinicians who perform augmentative communication evaluations become cognizant of the eye gaze behaviors of girls with RS and use this modality in planning interventions and communication systems. Until more research is available, clinicians will have to rely on dynamic assessments in multiple contexts to determine if an individual girl is using eye gaze primarily to point at objects or if she is capable of more complex mutual eye gaze behaviors (i.e., gaze switching). This may be a skill that can be developed with appropriate training and should be studied further.

The data are interpreted to conclude that the girls with RS who were reported on in this study were using multiple modalities to communicate and that the mean number was three modalities per girl. This finding also has implications for communication assessment and intervention planning. Clinicians should evaluate all modalities and consider the influence of apraxia, delayed response time, and inconsistency in responding when planning interventions. While examining the data and reading the text responses it became clear that girls may use different modalities based on multiple factors. Seizure activity and level of awareness were reported as factors that could influence quality of responsiveness in RS. An observation that was described repeatedly was that an individual girl's performance could vary greatly from day to day. It has been observed that girls with RS may not be always sufficiently alert to engage in optimal communicative interactions (Woodyatt, Marinac, Darnell, Sigafos, & Halle, 2004). Teachers are encouraged to allow time for their students with RS to achieve a sufficient state of alertness to participate fully in communicative interactions (Ryan et al., 2004). By providing a girl with RS

multiple modalities to communicate, and by training communication partners to recognize level of alertness and types of responses, the communicative and educational potential of these girls may be better realized. Outcome data on effectiveness of types of interventions in this population are clearly needed.

There were several limitations to this study. There was a sizeable difference in the number of respondents in each group, with far more parents responding than professionals. Any interpretation of group differences must weigh the disparate sizes of the groups. In addition, the respondents were self-selected, had Internet access to the survey, and were connected in some way to the IRSA. Because of their connections in the RS community, this group may therefore be uniquely sensitized to the communication skills of girls with RS and their responses may not be reflective of those of other parents or professionals. Other limitations were that the diagnoses of the girls could not be confirmed and because of the anonymity of respondents, responses could not be verified. Whether any respondents reported on the same girl with RS also could not be confirmed. The inability to collect data on the stage of RS was a clear limitation. Bartolotta (2004) indicated that most parents could reliably report which stage of RS a particular girl was in, but professionals were unable to report those data. It would have been helpful to be able to analyze the relationship between stage of RS and level of communication skills as part of this study. This should be explored in future research.

One-third of the respondents reported on the communication skills of girls in the 4 to 7 year age range. It is likely that parents and professionals who work with young, newly diagnosed children track new developments in treatment and therefore were most likely to respond to the invitation to participate in this study. Parents of older girls, who may be less likely to be searching for new treatments, are thus less likely to respond. The results of the study were heavily skewed toward behaviors of girls in the 4 to 7 year age range. Those in this group, likely to be composed of girls in Stage 3 of the disorder, are typically past the regression stage and in the period when they may be learning new skills or regaining skills previously lost. They also are likely to be enrolled in educational and therapeutic programs, where professionals may be exploring modalities for communication. This then may account for the overwhelmingly positive responses regarding the communication abilities of the girls in this study.

In this study, the perceptions of parents and professionals with regard to the communication skills of girls with RS were fairly similar. This observation can help strengthen the reliability of descriptions given by parents about their daughters though strategies to verify information are clearly warranted. What also is clear is that familiarity with an individual girl is important. As is common when interacting with

any individual with a severe disability, the communication partner must use inference and intuition to ascribe meaning to the person's behaviors (Grove et al., 1999; Sigafos, Drasgow, Reichle, O'Reilly, & Tait, 2004). This does not imply that the communication partner overestimates competence on the part of an individual with a disability. Rather, there is an assumption that competent behavior is possible, and then the communication partner must work to accurately interpret the signals. In this study, girls with RS were reported to be communicating with their parents, their teachers, and the SLPs who work with them. All of these individuals are working hard to untangle the meanings conveyed by the girls with RS and to accurately interpret their behaviors. There is much work to be done to develop clinical processes for accurate assessments that can be utilized in the presence of severe disability. We can use the perceptions of these familiar partners, combined with their rich descriptions of the behaviors they observe, to develop objective measures of skill level and outcomes of the intervention we provide to girls with RS.

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