



RETT SYNDROME Communication Guidelines

RETT SYNDROME Communication Guidelines:

A handbook for therapists, educators, and families



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Introduction, page 4: reference to “Appendix 6” was corrected to “Appendix 5”.

Section 4, page 27: “decreasing” was deleted in the heading “Fatigue, alertness, sensory regulation, stress and anxiety” so that the format was consistent with other headings in this section.

Section 8, page 51: “making” was corrected to “making choices” in the Communicative Functions text box.

Section 8, page 62: reference to “Appendix 2” was corrected to “Appendix 5”.

Appendix 1, pages 75, 79, 81-82, 84, 86-88: format was amended to aid clarity (line spacing and headings).

Appendix 3, pages 93-96: AAC Profile and ACETS were added; TASP and VB-MAPP were removed; and C-BiLLT entry was revised.

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Many thanks to everyone who has contributed to the development of these guidelines. This project has been a joint effort by many people over a number of years and a truly international collaboration, an example of the teamwork that is such a key element in communication assessment and intervention.

Developing language and communication is a lifelong process, and we hope the guidelines shared in this handbook will support individuals with Rett syndrome, their families, and the professionals working with them as they make that journey together.

Gillian Townend
Theresa Bartolotta
Anna Urbanowicz
Helena Wandin
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As the International Rett Syndrome Foundation (known today as Rettsyndrome.org), we are proud to be a part of bringing this global research initiative to life for families affected by Rett syndrome and the community that is committed to helping them achieve more. The work of Gillian Townend, Theresa Bartolotta, Anna Urbanowicz, Helena Wandin, Leopold Curfs, and many other researchers and contributors is an excellent representation of the dedication and commitment to improving the quality of life for our children and loved ones living with Rett syndrome. It is our honor to support these distinguished researchers. We are grateful for their work.

We remain unwavering in our commitment to empowering you today while working to transform your tomorrow.

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All websites listed in this book can be accessed online at communication.rettysyndrome.org.

Foreword

When she was a year old, she spoke her first few words. When she was diagnosed with Rett syndrome 18 months later, she had completely lost her ability to speak. How was she to communicate her wants, needs, feelings, and opinions now?

For many families living with Rett syndrome, finding a way to communicate is one of their most challenging issues. Learning how to use augmentative and alternative communication (AAC) and learning how to facilitate their loved ones to maximize their potential takes belief in their abilities, a great deal of effort, and lots of support. Seeking knowledge and skills on how to adjust assessments and how to manage communication, parents (and professionals) find themselves in need of expert support, in need of a shared vision of how to approach these aspects. Yet, so far, little evidence-based guidance has been available to them.

To do justice to the specific challenges of Rett syndrome, such as apraxia, breathing disorder, and epilepsy, a broad consensus on assessment, intervention, and long-term management of communication is needed – for research as well as daily communication and education. The collaboration between parents, caregivers, professionals, and experts in the field of communication for Rett makes these international guidelines the valuable document that is presented here. These guidelines are an important step forward in enabling people with Rett syndrome to communicate more effectively.

Many girls, now teenagers and adults, have moved on from learning to use a few pictos to using a combination of low tech and robust vocabulary on eye-gaze devices. Young girls and boys are starting out with eye-gaze-controlled communication systems. They are learning to communicate, to tell us their wants, needs, feelings, and opinions. It is exciting to see how new possibilities are helping to change our perspective on their cognitive abilities.

Many thanks to the project team, to all the committed communication specialists and parents and caregivers who contributed to these guidelines. Our beautiful daughters and sons with Rett syndrome may be unable to use their own voices to speak, but surely these guidelines will help them move forward in communicating their needs and opinions and in speaking their minds.

On behalf of all Rett parents everywhere,

Mariëlle van den Berg

Chair, Rett Syndrome Association Netherlands (Nederlandse Rett Syndroom Vereniging)

Introduction

Background

Rett syndrome is a neurodevelopmental disorder affecting approximately 1 in 10,000 females. It occurs rarely in males. It is often associated with severe physical and communication disabilities. In most cases, the cause of Rett syndrome is linked to a mutation in the methyl-CPG-binding protein 2 (MECP2) gene.

Why we developed the communication guidelines

A severe disruption in communication skills is one of the characteristic features of Rett syndrome. This has a fundamental impact on the quality of life for individuals with Rett syndrome. Although the literature on best practices in communication assessment and intervention in Rett syndrome is growing, caregivers around the world continue to report their struggle to access appropriate, knowledgeable, timely, and ongoing assessments, interventions, technology, support, and advice tailored to the specific communication needs of the person with Rett syndrome. Communication professionals also report challenges in finding the information, training, and support they need to build up their knowledge and expertise in this area. Therefore, we developed the communication guidelines presented in this handbook to help caregivers, communication professionals, and others support the communication development of individuals with Rett syndrome.

How we developed the communication guidelines

The idea of creating guidelines came into being when we, a small group of motivated professionals working in the field of Rett syndrome and communication, joined together following the 3rd European Rett Syndrome Conference in Maastricht, The Netherlands, in October 2013. With funding from Rettsyndrome.org, we devised and led a project to develop the Rett Syndrome Communication Guidelines.

The project involved reviewing the literature and conducting surveys to identify best practices in the assessment, intervention, and longer-term management of communication in individuals with Rett syndrome around the world. Around 650 people from 43 countries participated in the project, with 490 caregivers and 120 communication professionals completed the surveys, and 36 professionals and parents forming our expert panel. As the project team, we developed draft statements and recommendations based on the findings of the literature reviews and the survey responses from caregivers and professionals. The draft statements and recommendations were then reviewed twice by our expert panel. The panel provided feedback to the project team, and we revised the statements and recommendations until consensus was reached. The final set of statements and recommendations that reached consensus became the Communication Guidelines. This means that these guidelines are firmly based on findings from the available literature combined with expert opinions from professionals and caregivers around the world.

The purpose of this handbook

This handbook was designed to share the recommended guidelines for the assessment, intervention, and longer-term management of communication in individuals with Rett syndrome and to provide relevant background information for caregivers and professionals. These guidelines are intended for all individuals with a diagnosis of typical or atypical Rett syndrome, both male and female, wherever they were born and wherever they live. The guidelines were created to be adaptable to different situations and circumstances. They do not offer step-by-step instructions that people in all countries and services must follow and apply in an identical way. They aim to provide basic information that families around the world can share with therapists and educators to enable any individual with Rett syndrome to meet their full communication potential.

How to use this handbook

The handbook is divided into eight sections, with information and recommendations relating to the following: (1) guiding principles; (2) professional practice; (3) features of Rett syndrome and coexisting conditions; (4) strategies to optimize engagement; (5) general communication assessment; (6) AAC assessment; (7) assessment of AAC system/device; and (8) intervention.

The handbook can be used in many different ways. It can serve as a guide for designing assessments that reflect best practices and enable teams to develop goals that are functional and meaningful. It can provide strategies for families and therapists to advance the communication skills of any individual with Rett syndrome. The handbook provides information on how to get started with communication as well as how to progress the skills of more experienced communicators. There is also key information that describes how the unique features of Rett syndrome may impact communication. Individuals looking for information on specific topics can consult the Table of Contents at the front of the handbook. Other readers who are new to Rett syndrome may find that the early chapters provide a foundation of information to help them as they begin their work with an individual. The handbook can serve as a tool for educating people about Rett syndrome and for advocating for the communication services and support they need.

Throughout the book, the guidelines are presented in several formats. Some are shown in boxes that stand out from the text; others are embedded within the text. All are equally important. The ‘raw’ guidelines (the complete, final set of statements and recommendations that reached consensus) are presented as tables in Appendix 1 at the back of the book.

Quotes from the caregivers’ and professionals’ surveys and from the expert panel are included throughout the handbook to highlight important information. Photographs of individuals with Rett syndrome illustrate a broad range of communication systems and settings, and short stories offer “A Parent’s Perspective” on different stages of the communication journey. We are indebted to the many families who have graciously shared their photographs and their stories with us and are grateful to the even greater number of caregivers and professionals who took time to complete the surveys and engage with the project on multiple levels.

Links to a number of resources and useful websites and organizations are given in Appendix 5. This list is not intended to be exhaustive and will vary according to country and language. It is recommended, therefore, that each country produce its own list to supplement the Appendix. As far as possible, technical language is avoided in this handbook. Where this could not be avoided, the definition is usually given the first time a term is used. However, the Glossary of Terms on page 90 (Appendix 2) may also be helpful as you read through this book.

Language note:

1. In this handbook the term “individuals with Rett syndrome” is used. In some places this is shortened to “individuals” to make the sentences easier to read. Therefore, it is important to remember while reading the handbook that “individuals” always means “individuals with Rett syndrome.”
2. In some places the gender neutral “they,” “their,” or “them” is used, but in other places “she” or “her” is used. This is not intended to exclude males with Rett syndrome, and everything in this handbook should always be read as applying to both males and females with Rett syndrome.

STRATEGIES TO OPTIMIZE ENGAGEMENT



“ Make AAC available
at all times ”

Strategies to Optimize Engagement

Communication skills of individuals with Rett syndrome will fluctuate based on internal and external factors. Expect inconsistency.

Communication partners can use a number of strategies to initiate and maintain engagement with an individual with Rett syndrome. These strategies can be used in everyday life at home, school, work, and other settings. They can also be used during more specific communication assessment and intervention sessions. Different strategies may work better for some individuals than others and, even for one individual, at different moments in time. This may depend on factors such as their current mood, emotional state, quality of sleep, effects of breathing and respiratory abnormalities, epilepsy, medication, gastrointestinal issues, and dyspraxia/apraxia, as described in Section 3.

A PERSONAL COMMUNICATION PASSPORT¹ CAN BE...

an invaluable resource for sharing information about an individual's communication style, strategies that are most appropriate to use, and any personal/health-related factors that may affect their behavior and the choice of strategies/communication methods.

Attitudes and Behaviors of Communication Partners

Engagement Strategies

Address and talk directly to the individual with Rett syndrome.

When communicating with individuals with Rett syndrome in any situation, it is extremely important that communication partners address and talk directly to the individual with Rett syndrome rather than talking about them in their presence. This is extremely important for initiating and maintaining engagement. This may include not speaking over the individual with Rett syndrome or ignoring them in the presence of others. Sometimes communication partners might need to talk about the individual in front of them (e.g., a professional may need to ask a question the individual with Rett syndrome is unable to answer themselves). In these circumstances, the individual with Rett syndrome should still be included in the conversation. This might involve the professional explaining to the individual with Rett syndrome why they need to talk about them and/or asking the individual with Rett syndrome for confirmation of the information provided by their caregiver.

Engagement Strategies

Make eye contact and use multimodal communication.



“Always include her in any conversations. Never speak over her or ignore her when having a conversation with somebody else.”

“... make direct eye contact, move into her space slightly and be patient.”

Caregiver

“Communication should be multi-modal and change over time as all of ours does.”

“Follow the client's interests; use animated affect/voicing, playful obstruction.”

Expert panel

¹ For further information, see: Millar, S. & Aitken, S. (2003). Personal Communication Passports: Guidelines for Good Practice. *CALL Centre*. University of Edinburgh, Edinburgh. See also: <https://www.communicationpassports.org.uk/resources/>

It is also extremely important that communication partners make eye contact with the individual and use multimodal communication (facial expressions, gestures, vocalizations, speech alongside AAC) to attract and maintain the individual's attention. This may be accomplished, for example, by varying tone and intensity of voice, and using varied body movements, facial expressions, and gestures. Making eye contact can help the individual with Rett syndrome know their communication partner is talking to them and make it easier for them to pick up on non-verbal communication like facial expressions. It also helps the communication partner to perceive and respond to subtle communicative signals from the individual.

It should be noted, however, that some individuals may not like direct eye contact and may actively avoid it. In this case, other strategies need to be adapted to suit the individual's needs. Communication partners are not expected to use all modalities in every interaction but to be sensitive to the individual's preferences, needs, and mood, and to adapt their own behavior and interaction style accordingly.



In many cases, communication partners can maintain the individual with Rett syndrome's attention by being engaging, fun, and interesting. This includes making sure vocabulary, topics, and activities are interesting to the individual with Rett syndrome, keeping in mind that interests may or may not be age appropriate. Many caregivers and professionals report that music is an interest shared by most individuals with Rett syndrome. However, everyone is different, so it is important for communication partners to find out what is of interest to the specific individual and provide varied activities aligned with their interests.



“Communication is most successful if it starts with fun activities.”

“Putting all her favorite music as choices in both the PODD book and on the eye-gaze device has been the best way to motivate her to use the AAC.”

Caregiver

“Taking the time to understand how each individual uses natural communication skills, e.g., existing eye gaze and how it is used, yes/no responses.”

Professional

Engagement Strategies

Make sure vocabulary, topics, and activities are appropriate to the interests of the individual with Rett syndrome.

It is extremely important that communication partners spend time getting to know the individual with Rett syndrome in order to build a relationship. This involves being patient and persistent; it may take time to develop trust between an individual and a new communication partner. This is a vital step for development of communication skills and is especially important to keep in mind when gains in communication can be slow.

Communication partners should:

- be responsive in their interactions, acknowledging and reinforcing all communication attempts;
- follow the individual's lead;
- give feedback and attribute meaning (saying aloud what they think the individual's response/action/behavior means); and
- be consistent (in approach, language, and vocabulary used).

Consistently responding, acknowledging, and reinforcing all communication attempts helps individuals with Rett syndrome develop and maintain their communication abilities. It is very common that communication between individuals who rely on AAC and their speaking communication partners is unbalanced, with the communication partner often being the one to direct the interaction. By following the individual's interests and focus and providing wait time, the individual with Rett syndrome can be given opportunities to initiate communication, and the communication partner can follow the individual's lead. This may involve setting up activities of interest that allow the individual to take a lead and go at their own pace.

Engagement Strategies

Providing access to AAC systems and devices increases the individual's possibilities to initiate and take control/direct conversations more clearly.

Interaction with peers can also be invaluable in encouraging communication. This includes having other AAC users and neurotypical peers who can engage in age-appropriate topics and activities and can use their own or the individual's AAC system/device with them.



A PARENT'S PERSPECTIVE

Being around Sofia, another AAC user with Rett syndrome who is older than Tilly, was incredible in improving Tilly's communication. Having a positive AAC role model meant Tilly became more vocal and used her device and low-tech communication more. She even used Sofia's eye gaze spontaneously, too, to talk about her physiotherapy. Both Sofia's neurotypical sister as well as Tilly's neurotypical friends at nursery, using Tilly's device and interacting with her using AAC, have also been essential in normalizing AAC and helping Tilly to feel more confident using it. The introduction and nursery pages to help Tilly say their names and say what she wants to do have also been important at nursery.

Giving feedback and attributing meaning to the behavior of individuals with Rett syndrome, even if the meaning seems unclear, can also help to shape an individual's communicative behaviors and develop consistency of use. For example: "You looked at X and smiled; I think you are telling me you like X." "You looked at X and then at the word LIKE; I think you are telling me you like X." "You closed your mouth and turned away; I think you are telling me you don't want more Y." In this way, even if the communication partner's interpretation is wrong, the individual should begin to realize the connection between their behavior and the interpretation.

Engagement Strategies

Adapt natural situations and activities so that opportunities for communication are created.

If the individual with Rett syndrome uses one or more AAC systems or devices, it is extremely important that they are available at all times to enable the individual to make the most of every opportunity for communication. Opportunities to use their AAC system/device should be provided throughout the day and incorporated into all situations and activities involving the individual, such as when bathing, eating, changing clothes, and shopping. Words like "more," "finished," "like," and "don't like" can be used frequently throughout the day. Providing opportunities helps build vocabulary and language.



"In all communication there is interpretation, say your interpretation out loud and repeat it, then the child will know whether you have the same understanding."

Caregiver

"Acknowledge any attempt at communication."

"[Provide] feedback immediately, e.g., this is what I am seeing and this is what I think it means."

Professional



Engagement Strategies Make the individual’s AAC system(s) or device(s) available at all times.

For more examples, see *Building Foundational Communication Support for Individuals with Rett Syndrome*² from Rett University.

Engagement Strategies Keep communication in the ‘here and now.’

Keeping communication in the here and now (i.e., talking about current thoughts, feelings, and activities) allows for a stronger link between words and their meaning. This may be especially beneficial, for example, when learning new vocabulary.

Individuals with Rett syndrome often have better receptive language abilities than expressive. Although it may sometimes appear that they do not understand, it is important to explain what you are doing now and what you will be doing next, and to let them know when you will be changing and/or moving on to another activity. For example, use visual supports to indicate that an activity is ending and a new one will begin. This may be particularly helpful for individuals who have high levels of anxiety.

Strategies Targeting Internal Factors That May Impact Communication

Movement disorders

Containing stereotypies with an embrace or placing a hand over the individual’s hand(s) has been shown to help to focus attention and learning. Gently restraining the non-dominant hand or arm using a hand wrap or elbow splint may also be helpful. It is important, however, to refer to the relevant restraint policies and procedures in the country, location, or organization in which the individual resides and/or receives therapy or education before using any restraints. Seek advice from an OT.

Voluntary control over, and coordination of, motor movements requires a lot of physical and cognitive effort that can be taxing for individuals with Rett syndrome. Therefore, it is recommended that the motor demands of a task or activity are decreased as the cognitive load increases. For example, limiting the number of options on a page or use partner-assisted scanning (see page 58) for educational tasks.

Engagement Strategies Decrease motor demands as the cognitive load of a task increases.

“Access to a system in all aspects of life (school, home, community) where the entire team is on board with implementing the same system.”

“Use and train the communication in an everyday context (not an isolated context).”

Professional

“She needs access to AAC 100% of the time.”

“...attempt to get the communication to be a natural part of the everyday life.”

Caregiver

“Need multiple opportunities to practice all modes across their day to increase communication abilities.”

Expert panel

“If the student has extremely limited physical abilities for direct selection, balance the physical effort of communication against the physical/mental effort for academic learning/ responding vs the physical effort to activate environmental controls.”

Professional

² Rett University, (n.d.). *Building Foundational Communication Support for Individuals with Rett Syndrome*. <https://rettuniversity.org/wp-content/uploads/2015/05/RettU-Communication-One-Page-FINAL.pdf>

Breathing/respiration irregularities

Observe the individual closely for episodes of breath holding and hyperventilation, and wait for these to pass. Seek medical help and advice when necessary.

Epilepsy

Be vigilant and observe for any signs of a seizure. Wait for it to pass. Seek medical help and advice when necessary.

Scoliosis/kyphosis

Make sure the individual is well supported in whatever position they are in (e.g., if seated, make sure they have a supportive system if required). Observe for signs of pain or discomfort. Consult with an OT, PT, and/or orthopedic consultant. You can also refer to the Scoliosis in Rett Syndrome booklet³ produced by the Telethon Kids Institute.

Fatigue, alertness, sensory regulation, stress and anxiety

Ask the individual or their caregivers about their sleep patterns. Observe for signs of fatigue. If the individual is tired, keep activities short or do things that do not require a lot of concentration. Be aware, however, that for some individuals falling asleep may be a sign of understimulation, and they may need more stimulation/activity to increase their alertness.

Engagement Strategies

Individuals may need help to regulate their levels of alertness and their readiness to participate in daily living and educational activities.

Some individuals may, for example, find it easier to engage while they are standing or after a short break during which they were able to move around. Others may find that short bursts of watching television/films or listening to music help to regulate their body. Different sorts of music can have different effects (calming or stimulating) and can be used at different times for different purposes.

Engagement Strategies

Pay attention to behaviors that indicate the need for a change in position or a change/break in activity.

It is important to observe the individual's preferred positions for different activities and to look for signs that they need a change in position or in sensory stimulation, or a break/change in activity. Ask if they would like a change or a break and, if necessary, assist them in changing position or type of sensory stimulation, and/or provide them with a change of activity. It is important to incorporate words and phrases into an individual's AAC system that allow them to ask for a break or change of position/activity. These words and phrases should be easy to access and should be modeled to demonstrate their use.

Individuals with Rett syndrome may show signs of stress/anxiety when there is either (a) excessive stimulation and a high level of demand or (b) a lack of stimulation and/or demand. It is important to consider how to maintain a balance between activating/stimulating activities and breaks in activity. Providing the individual with a high level of control can offer protection from stress and anxiety.

A high level of control may be achieved when the individual:

- understands and is prepared for what is happening,
- has means to express themselves and is listened to, and
- can influence what is happening.

³ Downs, J., Bergman, A., Carter, P., et al. (2009). *Scoliosis in Rett Syndrome: A Collaboration Between Parents, Clinicians and Researchers*. Telethon Institute for Child Health Research. Perth, Western Australia. https://rett.telethonkids.org.au/siteassets/media-docs---rett-syndrome/scoliosis_booklet.pdf

Signs of the need to increase alertness or sensory input/change activity include:

- falling asleep/drowsiness and
- inattentiveness.

Suggested strategies include:

- increasing the intensity of your interaction (e.g., increasing the loudness of your voice and becoming more animated);
- changing position (e.g., getting everyone to stand up if you have been seated so that the individual can move from sitting-to-standing);
- using sensory input (e.g., vibration, music, and movements); and
- changing activity.

Signs of the need to decrease sensory input/change to a “recovery activity” include:

- grimacing or complaining, and restless motor activity;
- tensing of the body;
- getting up/walking away from a situation; and
- changes in breathing (e.g., hyperventilation and breath holding).

There are individuals with Rett syndrome who experience respiratory changes that are not under their control, so they may hyperventilate or breath hold regardless of the situation they are in. However, changes in breathing may occur with substantial sensory input, when the individual is excited, or when experiences demands are high. Interviews with caregivers and communication partners can provide insight into the impact of stress on the respiratory behavior of an individual.

Suggested strategies include:

- decreasing the intensity of the interaction (e.g., decreasing the loudness of your voice);
- providing a break;
- decreasing demands; and
- providing calming tactile input (e.g., squeezing shoulders, arms, hugging the person tightly, and/or placing a weighted blanket or pillow on the person’s lap).

REMEMBER THAT EACH PERSON IS DIFFERENT AND WHAT CALMS ONE PERSON MAY AGITATE ANOTHER.

These suggestions are general strategies – make sure you talk to the person and other people who know them well about what works for them. The demands of a particular situation may also be too high or too low for the individual, which then can result in increased stress. Communication partners should learn about the individual with Rett syndrome and create an atmosphere that is supportive yet challenging.

Suggested strategies to reduce anxiety include:

- asking open-ended questions that don’t have a right or wrong answer,
- giving a high chance of success by minimizing any pressure on responding, and
- modeling – not testing.



“The higher the cognitive demand, the lower the physical demand needs to be.”

Professional

“She fatigues and requires periodic breaks.”

Caregiver

“Breaks should be determined according to the girl’s response and attention at the time.”

Expert panel

The “traffic light system” developed by Judy Lariviere can be helpful for training caregivers to recognize levels of sensory arousal and to develop the strategies needed to adjust them. It also helps individuals with Rett syndrome to recognize their own internal state. The Sensory Regulation Chart⁴ (see below) provides a color-coded version of the various levels of sensory regulation based on how an individual is processing sensory input in their environment. It includes a description of some of the strategies that can be used to support someone in returning to Level 1 (functional state of arousal) so that they are in an optimal condition (ready) to communicate and to learn.

FIGURE 4: SENSORY REGULATION CHART

OVERSTIMULATED Agitated/sensory overload	Level 3	<ul style="list-style-type: none"> • Needs strong sensory input from sensory diet to assist in calming down; overwhelmed by sensory input. • Will take longer to return to Level 1 when sensory system moves into Level 3 or “red zone.”
OVER RESPONSIVE Needs to be “calmed down”	Level 2	<ul style="list-style-type: none"> • Needs sensory input or a sensory break based on sensory diet to help return to Level 1; snack or drink; music can help re-engage; change of activity; movement and/or change of position
FUNCTIONAL STATE OF AROUSAL “Regulated state”	Level 1	<ul style="list-style-type: none"> • Ready to learn, communicate, and actively participate • Calm; body supported and relaxed • Sitting upright and/or remaining seated • Establishing eye contact; smiling; giggling • Interactive; easily looking between different things • Communicating with communication partner(s) • Engaged, playing, having fun • Does not seem anxious or tired
UNDER RESPONSIVE Needs to be “revved up”	Level 2	<ul style="list-style-type: none"> • Needs sensory input based on sensory diet to help return to Level 1; typically needs a break involving movement/change of position or snack to re-energize and/or listening to favorite music; change of activity.
UNDERSTIMULATED Falling asleep/shutdown	Level 3	<ul style="list-style-type: none"> • Needs strong sensory input from sensory diet with extended break to assist in “waking up” or “revving up” sensory system.

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⁴ For further information, see: Lariviere, J. (2015). Eye-Gaze Technology for Girls with Rett Syndrome: From Trials to Conversations. *Closing the Gap Solutions*. December 2014/January 2015, 4-13.



Auditory processing difficulties

Referral to an audiologist may be helpful in understanding how an individual perceives and processes auditory stimuli.

If an individual appears to have difficulties functioning with high levels of background noise, reducing unnecessary auditory input may help (e.g., turning off the radio/TV).

Visual impairments

Referral to an eye specialist, such as an ophthalmologist or optometrist, may be helpful in understanding how the individual perceives and processes visual stimuli.

For some individuals it may help to increase the size of symbols, simplify the complexity, use high-contrast symbols, or arrange the display so that the symbols are placed to one side (left or right), laid flat on a horizontal surface, or arranged vertically. Others may benefit from a reduction in visual distractions (e.g., placing the individual’s AAC device directly in front of them while moving other items outside of their visual field).



“Some individuals need distraction-reduced settings to show true communication potential.”

Expert panel

Additional Considerations

Structured settings

Engagement Strategies	Reduce distractions.
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In a more structured setting — i.e., in an assessment or intervention (therapy or teaching) session — it is very important to reduce distractions. Some individuals with Rett syndrome will find it harder to concentrate in a noisy and busy environment. Reducing distractions, particularly when learning a new skill, can initially help the individual focus, attend, and learn. Reducing distractions not only benefits the individual, but also the communication partner who is better able to focus and attend to communicating with the individual. However, so as not to limit the individual, it is important that they also get used to communicating in a more typical environment (i.e., with higher levels of background noise or visual stimuli). As with motor challenges, there must be a balance of sensory and cognitive demands.

Response time

Engagement Strategies

Be aware of delayed response time.

Individuals with Rett syndrome may have a delayed response in communication interactions. Response delay may be for a few seconds or up to a minute or more. This can vary from day to day and throughout the day, depending on a wide range of factors, such as the presence of dyspraxia/apraxia and levels of motivation and fatigue. Therefore, a slow response or an absent response is not necessarily an indication of a lack of understanding.

To determine sufficient wait time for a particular individual, communication partners can do the following:

- Observe the individual to identify typical communication behavior.
- Review videos of communication interactions to identify typical communication behavior.
- Discuss typical communication behavior with familiar communication partners.
- Consider how the features of Rett syndrome and any coexisting conditions may impact response time.



“The answering time can sometimes be extra-long and sometimes extra quick.”

“She has apraxia and may take a while to respond.”

Caregiver

“Sufficient time to wait will vary from individual to individual and this should be identified following observational assessment.”

Expert panel

