RETT SYNDROME
Communication Guidelines

Netherlands
RETT
EXPERTISE CENTRE
Maastricht University Medical Centre

Rettsyndrome.org
ACCELERATING RESEARCH. EMPOWERING FAMILIES.

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Communication Guidelines:
A handbook for therapists, educators, and families

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Erratum: December 2020
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.

Acknowledgment from Authors

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
Leopold Curfs

February 2020

Acknowledgment from Rettsyndrome.org

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 honour 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.

Table of Contents

All websites listed in this book can be accessed online at communication.rettsyndrome.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 maximise 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 completing 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 optimise 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 organisations 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.
3. In this UK and Ireland edition spellings have been changed from the original American English to British English. The terms "speech-language pathologist" and "physical therapist" have also been changed to "speech and language therapist" and "physiotherapist" (except where these related to named people's job titles in Appendix 6) to reflect common usage in the UK.

AAC ASSESSMENT

> “No prerequisite skills must be demonstrated”

SECTION 6

AAC Assessment

AAC may be unaided or aided.

Unaided AAC means not using external tools to communicate but relying on the user's own body to convey messages. Examples include gestures, signs, or vocalisations.

Aided AAC means using external tools, devices, or systems to enhance communication. These may be electronic or non-electronic (high-tech or low-tech).

Aided AAC includes a range of communication methods, from using a small number of objects or pictures for choice making, to robust language systems with a large number of symbols. These pictures and symbols can be presented on a single page, in a multi-page communication book, or on an electronic device such as a computer, tablet, or a dedicated device that is only used for communication.

A low-tech AAC system is not electronic and does not require a power source (e.g., individual objects, photos, and communication board/book).

A high-tech AAC system is an electronic device with speech output. The screen can have a static or a dynamic display, and the output can be recorded/digitised speech or synthesised speech. High-tech systems range from simple single message devices to complex computer-based systems.

A speech generating device (SGD) is a type of high-tech device, usually used to refer to a more complex computer-based system rather than a simple single-message device.

Both low- and high-tech systems can be accessed a number of ways, including direct touch or eye gaze.

An eye-gaze device (eye-gaze technology) is a type of SGD that is controlled by eye gaze. The device has either a built-in eye tracker or an eye-tracker module that connects to the device to detect and track where the user is looking. Usually, the speed and accuracy of selection can be adjusted to the needs of each user—e.g., the length of time the user must look at an item on screen in order to select it (dwell time) and the size/number of items on the screen.

> "The very first barrier that needs to be taken away in many situations is the lack of sufficient AAC due to the expectations of the professionals (SLT, teacher). All doctors that are informing parents about the diagnoses should direct them to AAC and aided language and their opportunities and NOT mention anything about cognitive disabilities."
>
> *Professional*

> "AAC can be introduced for children or adults who are pre-intentional and have no means to demonstrate "cause and effect" cognition. AAC can support the development of these skills."
>
> *Expert panel*

Starting Point - Assessment of Readiness for AAC

AAC Assessment	No prerequisite skills must be demonstrated before aided AAC should be considered.

According to current practices, no prerequisite skills must be demonstrated before aided AAC should be considered. Aided AAC can be an option even before an individual has demonstrated understanding of cause and effect, clear choice making, or communicative intent. Aided AAC can assist development of these skills. Therefore, individuals should be referred for AAC evaluation at the time of diagnosis or shortly thereafter in order to maximise their communicative and linguistic development and to prevent negative effects, such as learned helplessness, that can arise from a lack of communicative opportunities.

Components of AAC Assessment

Any AAC assessment should consider multiple modalities, both unaided (facial expression, gestures, vocalisations) and aided communication (low-tech and high-tech options, depending on availability).

AAC Assessment	AAC assessment should include consideration of: • a range of symbol systems, • layout (e.g., size and number of options/pictures on the communication board or screen), and • complexity of vocabulary on offer.

Although the availability of certain symbol systems will vary according to country and language, the number and range of systems available around the globe is growing rapidly. Some symbols may be more suited to one individual than another (e.g., high-contrast symbols designed specifically for people with visual difficulties). Individuals may also have their own personal preferences that need to be taken into account.

In addition, attention must be paid to the layout and complexity of vocabulary during an AAC assessment. Issues relating to organisation of vocabulary will be explored further in Section 8.

> “Try as much software as possible and then begin with the most suitable one.”
>
> *Caregiver*
>
> “When reviewing symbol systems, it is important to take into consideration preferences and accessibility.”
>
> *Expert panel*

AAC Assessment	AAC assessment should include consideration of: • options for alternative access (e.g., eye gaze, touchscreen, and switch control), and • positioning (of individual and device, and AAC system).

Assessment should include identification of the most appropriate access method for an individual (e.g., eyes and hands). This will influence their potential to access and use the vocabulary on offer to them. How the device and the individual are positioned can be crucial to their success.

Models of Best Practice for AAC Assessment

AAC Assessment

There are a number of models of best practice in AAC that are appropriate to use with individuals with Rett syndrome. These include:

- the Six-Step Process,
- the Participation Model, and
- the Model of Communicative Competence.

The Six-Step Process (Dietz et al, 2012)[1]

This model follows a process similar to that of Dynamic Assessment. It suggests that AAC assessment consists of a series of steps, with the entry point determined by an individual's needs:

- Assessing communication using scenarios
- Considering the need for alternative access
- Incorporating multiple modalities (e.g., low-tech options)
- Providing AAC instruction
- Assessing a range of symbol systems
- Arranging device trials

The Participation Model (Beukelman & Mirenda, 2013)[2]

In this model, participation is central. A Dynamic Assessment approach is used to assess an individual's participation according to their opportunity and access barriers. The needs of the communication partners are also considered. The steps include the following:

- Identifying how the individual currently participates
- Assessing barriers to participation, including the potential to use AAC systems and/or devices
- Planning and implementing interventions, including provision of instruction to both the individual and their communication partners
- Evaluating how effective the intervention is, i.e., whether the individual's participation is increased
- If need be, the assessment starts again and the cycle repeats.

The Model of Communicative Competence (Light & McNaughton, 2014)[3]

In this framework, four specific areas of skill are assessed, together with two broader factors. Each of these is important for the development of an individual AAC user's communicative competence and forms a starting point for the identification of targets for intervention.

These are the four skills:

- Linguistic
- Social
- Operational
- Strategic

These are the other two factors:

- Psychosocial (internal to the individual – motivation, attitude, confidence, and resilience)
- Environmental (external to the individual – policy, practice, knowledge, attitude, and skills)

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[1] For further information, see: The Six-Step Process in: Dietz, A., Quach, W., Lund, S. K., & McKelvey, M. (2012). AAC assessment and clinical-decision making: the impact of experience. *Augmentative and Alternative Communication.* 28(3), 148-159.

[2] For further information, see: The Participation Model in: Beukelman, D. R., & Mirenda, P. (2013). *Augmentative and alternative communication: supporting children and adults with complex communication needs.* Brookes. Baltimore, MD.

[3] For further information, see: The Model of Communicative Competence in: Light, J. & McNaughton, D. (2014). *Communicative competence for individuals who require augmentative and alternative communication: a new definition for a new era of communication?* Augmentative and Alternative Communication. 30(1), 1-18.

Assessment of Readiness for Eye-Gaze Technology

Overall health, oculomotor skills, visual attention and memory, motivation, ability to focus on the screen, and prior experience with aided AAC will all impact ability to access eye-gaze technology. A careful and comprehensive assessment of these features can help to determine whether an individual is a strong candidate for eye gaze, if they will benefit from additional investigation, if they need a trial of an alternative access method, or perhaps if a combination of eye-gaze technology and other access methods are needed.

> “Some people with very complex health issues use eye-gaze very successfully; if you wait for someone to be well, it will never happen!”
>
> *Expert panel*

Readiness for eye-gaze technology	Assessments of readiness to use eye-gaze technology are best conducted through informal activities.

Assessment activities should be personalised as far as possible (e.g., introducing photographs of familiar people into on-screen activities or using motivating activities in the assessment). This may enhance the individual’s engagement. There are currently no formal assessments that reliably predict how the individual will communicate using eye-gaze technology. In accordance with the principles of Dynamic Assessment, the same activities used to assess readiness can also be used to train/build the skills that are necessary for use of eye-gaze technology for communication.

Readiness for eye-gaze technology	An individual does not have to ‘pass’ assessment at an early level of screen use (e.g., demonstrate cause-effect understanding) before their response to more advanced levels of eye-gaze technology use can be assessed (e.g., introducing them to grids for communication).

If an individual does not seem to be interested or motivated to engage with eye-gaze technology during the assessment, it should not be discounted as a possible access method. Lack of interest or motivation may be due to lack of experience on the part of the individual (i.e., they did not know what to expect from the activity/device or the activities used in the assessment were not sufficiently challenging/stimulating or interesting for the individual). Prior experience of picture communication and exposure to engaging on-screen activities may increase the individual’s engagement with eye-gaze technology. For some individuals, using communication grids to interact/converse with a communication partner may be more motivating than, for example, simple cause-effect games designed to practice controlling eye-gaze technology.

Readiness for eye-gaze technology	It is not necessary to achieve successful calibration on an eye-gaze device in order to assess eye gaze as an access method.

Some individuals may not be able to achieve a good calibration immediately. Initially, a default calibration or a parent's calibration may work well enough. It is also possible to do a simple one-point calibration or to adjust the calibration settings in other ways. Interactive games, if the individual finds them interesting, can be used to establish calibration of eye gaze over time (e.g., by providing experience of 'chasing' a target with the eyes). Any use of on-screen activities, including the use of communication applications, may increase the chances of a successful calibration at a later stage of the assessment process.

Readiness for eye-gaze technology	It is valuable to track and record an individual's eye movements and on-screen gaze pattern during assessments for eye-gaze technology.

> "It is not essential to obtain a good calibration and one should not spend time on this, especially in the initial stages as it could prevent moving on to other more important tasks. When the need for more accurate access becomes apparent, e.g., smaller and more symbols per page, then better calibration is required."
>
> *Expert Panel*

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