



RETT SYNDROME Communication Guidelines

RETT SYNDROME Communication Guidelines:

A handbook for therapists, educators, and families



This UK and Ireland edition is printed with support
from Rett UK



This project was financed by a HeART Grant from Rettsyndrome.org.

The international core work group was coordinated by the Rett Expertise Centre Netherlands-GKC.

Members of this group (the project team) were:

- **Gillian S. Townend, PhD CertMRCSLT**, Speech and Language Therapist and Researcher, Rett Expertise Centre Netherlands-GKC, Maastricht University Medical Center, Maastricht, The Netherlands.
- **Theresa E. Bartolotta, PhD CCC-SLP**, Speech-Language Pathologist and Adjunct Professor, Department of Speech-Language Pathology, School of Education, Monmouth University, NJ, USA.
- **Anna Urbanowicz, PhD**, Occupational Therapist and VC Postdoctoral Fellow, Social and Global Studies Centre, School of Global, Urban, and Social Studies, RMIT University, Melbourne, Australia.
- **Helena Wandin, PhD reg. SLP**, Speech-Language Pathologist, Swedish National Center for Rett Syndrome and Related Disorders, Frösön, Sweden.
- **Leopold M. G. Curfs**, Director of the Rett Expertise Centre Netherlands-GKC, Maastricht University Medical Center, and Professor of Intellectual Disabilities, Maastricht University, Maastricht, The Netherlands.

© Rett Expertise Centre Netherlands-GKC / Rettsyndrome.org

1st Edition: 2020

The material in this handbook is subject to copyright. It may be used and distributed freely as long as the authors are acknowledged. Translation into other languages is encouraged; however, permission to translate must be obtained from the authors prior to translation.

Contact: Gillian Townend, g.townend@maastrichtuniversity.nl

Journal reference for the guidelines:

Townend, G.S., Bartolotta, T.E., Urbanowicz, A., Wandin, H., Curfs, L.M.G. (2020). Development of consensus-based guidelines for managing communication in individuals with Rett syndrome. *Alternative and Augmentative Communication*, 36(2), 71-81.

Citation for this book:

Townend, G.S., Bartolotta, T.E., Urbanowicz, A., Wandin, H., Curfs, L.M.G. (2020). Rett syndrome communication guidelines: a handbook for therapists, educators and families. Rett Expertise Centre Netherlands-GKC, Maastricht, NL, and Rettsyndrome.org, Cincinnati, OH.

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

Foreword	2	Section 7: Assessment of AAC System/Device	46
Introduction	3	Trial Periods as Assessment for an AAC System or Device	47
Section 1: Guiding Principles	6	Assessment of Device-Specific Features	48
Rights of the Individual with Rett Syndrome	7	Section 8: Intervention	50
Beliefs and Attitudes.....	10	General Principles for Intervention	51
Section 2: Professional Practice	12	Developing Goals for Intervention	54
Principles of Teamwork	13	Targets and Goals for Intervention.....	55
Responsibilities of Professionals.....	14	Communication Functions	58
Rett Syndrome Specialist Clinics and Expertise Centres	15	AAC Systems.....	58
Section 3: Features of Rett Syndrome and Coexisting Conditions	16	Developing Vocabulary.....	62
Features Associated with Main or Supportive Clinical Criteria.....	18	Choosing and Organising Vocabulary	63
Coexisting Conditions.....	20	Access Methods.....	66
Section 4: Strategies to Optimise Engagement	22	Reading and Writing.....	70
Attitudes and Behaviours of Communication Partners.....	23	A Final Word on Intervention Techniques.....	72
Strategies Targeting Internal Factors that May Impact Communication.....	26	Appendices	74
Additional Considerations	30	Appendix 1: Statements and Recommendations	75
Section 5: Assessment	32	Appendix 2: Glossary of Terms	90
General Principles of Assessment.....	33	Appendix 3: Resources for Assessment of Communication	93
Assessment of the Individual	35	Appendix 4: Resources for Development and Tracking of Communication Goals	97
Assessment Procedures.....	36	Appendix 5: Links to Useful Websites and Organisations	98
Standardised Assessment or Dynamic Assessment?.....	36	Appendix 6: Advisory Group and Expert Panel Members	102
Other Considerations for Assessment.....	39		
Section 6: AAC Assessment	40		
Starting Point - Assessment of Readiness for AAC	42		
Components of AAC Assessment.....	42		
Models of Best Practice for AAC Assessment	43		
Assessment of Readiness for Eye-Gaze Technology	44		

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

GUIDING PRINCIPLES



“Communication partners should have an open-mind to the communication potential of the individual”

Guiding Principles

Rights of the Individual with Rett Syndrome

The United Nations (UN) Convention on the Rights of Persons with Disabilities¹ is a human rights document that declares that all persons with all types of disabilities must enjoy all human rights and fundamental freedoms.

The table below outlines the communication rights of individuals with Rett syndrome that were developed as part of the guidelines project. They are based on the UN Convention and were strongly endorsed by the expert panel. These communication rights underpin the guidelines, and the rest of the handbook explores what they mean in greater depth.

Rights of the Individual

In accordance with the UN Convention on the Rights of Persons with Disabilities, all individuals with Rett syndrome should:

- be treated with respect;
- have access to a comprehensive multidisciplinary assessment of their strengths and needs;
- have an appropriate communication system;
- have appropriate communication goals;
- have timely reviews and modifications of goals in line with changing needs;
- have access to advice, support, and services that start early and continue throughout life;
- have advice and support from knowledgeable and expert communication professionals;
- have communication partners who are trained in appropriate communication strategies and techniques;
- be offered activities appropriate to their age, interests, and culture;
- have the opportunity to make choices;
- be enabled to participate in society; and
- have the right to education.

Communication partners* should strive to uphold the communication rights of individuals with Rett syndrome. However, their ability to do so may be dependent on the resources that are available. For example, individuals living in rural or remote areas may not have access to a comprehensive multidisciplinary assessment of their strengths and needs, or the most appropriate communication system may be too expensive for them to purchase and may not be funded by the state or healthcare insurance. In addition, communication partners themselves may not be experienced in using the specific techniques or equipment needed to enhance and facilitate an individual's communication skills and will need some level of training if they are not already skilled. Yet, even in these situations, the basic rights outlined in the table should be aimed for as far as possible.

There are a number of similar documents that communication partners may also find helpful to share with others, including those responsible for policy/service development and planning, when discussing the communication rights of individuals with Rett syndrome. These include: **(a)** The Communication Bill of Rights developed by the United States National Joint Committee for the Communication Needs of Persons with Severe Disabilities (NJC).² It states that all people with a disability (to any extent or severity) have a basic right to influence, through communication, the conditions of their existence, and it outlines 15 specific communication rights that should be recognised and upheld in daily interactions. **(b)** A simplified and more accessible version of the Communication Bill of Rights created by SCOPE Australia. This can be particularly helpful when talking directly to individuals about their rights.

**Communication partner refers to anyone who interacts with an individual with Rett syndrome.*



“They have a right to make choices, and that should be part of a much bigger perspective on communication. They should have much more opportunities than just giving answers to choices.”


Expert panel

¹ For further information, see: <https://www.un.org/development/desa/disabilities/convention-on-the-rights-of-persons-with-disabilities.html>

² For further information, see: Brady, N.C., Bruce, S., Goldman, A., et al. (2016). Communication services and supports for individuals with severe disabilities: Guidance for assessment and intervention. *American Journal on Intellectual and Developmental Disabilities*. 121(2), 121-138.

The Communication Bill of Rights is a valuable resource for promoting communication as a basic right.

FIGURE 1: NJC COMMUNICATION BILL OF RIGHTS³



National Joint Committee for the Communication Needs of Persons With Severe Disabilities (NJC)

COMMUNICATION BILL OF RIGHTS

All people with a disability of any extent or severity have a basic right to affect, through communication, the conditions of their existence. Beyond this general right, a number of specific communication rights should be ensured in all daily interactions and interventions involving persons who have severe disabilities. To participate fully in communication interactions, each person has these fundamental communication rights:

1. The right to interact socially, maintain social closeness, and build relationships
2. The right to request desired objects, actions, events, and people
3. The right to refuse or reject undesired objects, actions, events, or choices
4. The right to express personal preferences and feelings
5. The right to make choices from meaningful alternatives
6. The right to make comments and share opinions
7. The right to ask for and give information, including information about changes in routine and environment
8. The right to be informed about people and events in one's life
9. The right to access interventions and supports that improve communication
10. The right to have communication acts acknowledged and responded to even when the desired outcome cannot be realized
11. The right to have access to functioning AAC (augmentative and alternative communication) and other AT (assistive technology) services and devices at all times
12. The right to access environmental contexts, interactions, and opportunities that promote participation as full communication partners with other people, including peers
13. The right to be treated with dignity and addressed with respect and courtesy
14. The right to be addressed directly and not be spoken for or talked about in the third person while present
15. The right to have clear, meaningful, and culturally and linguistically appropriate communications

For more information, go to the NJC website at: www.asha.org/njc

Brady, N. C., Bruce, S., Goldman, A., Erickson, K., Mineo, B., Ogletree, B. T., Paul, D., Ronski, M., Sevcik, R., Siegel, E., Schoonover, J., Snell, M., Sylvester, L., & Wilkinson, K. (2016). Communication services and supports for individuals with severe disabilities: Guidance for assessment and intervention. *American Journal on Intellectual and Developmental Disabilities*, 121(2), 121-138.

11220

Reprinted with permission.

³This printable version of the NJC Communication Bill of Rights is available at: <https://www.asha.org/uploadedFiles/NJC-Communication-Bill-Rights.pdf>

FIGURE 2: SCOPE AUSTRALIA COMMUNICATION BILL OF RIGHTS⁴



Reprinted with permission.

⁴ This printable version of the SCOPE Australia Communication Bill of Rights is available at: https://www.scopeaust.org.au/wp-content/uploads/2017/01/Bill-of-rights-2017_a4.pdf



Beliefs and Attitudes

Beliefs and Attitudes

The potential to communicate is frequently underestimated in individuals with Rett syndrome.

Individuals with Rett syndrome usually have better receptive language (understanding) than expressive ability. Caregivers and communication professionals frequently report that individuals with Rett syndrome know more than they are able to express or demonstrate. This is often due to the complex conditions that coexist with Rett syndrome, making it difficult for individuals to use speech, gestures, and other conventional ways to communicate. Section 3 has more detailed information regarding features of Rett syndrome and coexisting conditions that influence communication.

Beliefs and Attitudes

It is extremely important that communication partners have an open mind to the communication potential of the individual with Rett syndrome.

All communication partners should believe that individuals are capable of communicating and that, given the opportunity, they will be able to communicate with support from augmentative and alternative communication (AAC). Belief in each individual's ability to understand and potential to communicate is central when developing goals for communication and setting targets to improve communication skills. Caregivers and communication professionals often report that lack of belief in an individual's potential can be demotivating and have a negative effect on the individual. This does not mean that everyone should be expected to achieve the same level of complex communication, but everyone should at least be given opportunities to develop their skills as far as possible. Being open to possibilities is a key starting point.



“Believe that she is fully capable of communication if given the right circumstances!”

Caregiver

“The assumption that girls and women with Rett syndrome are severely or profoundly cognitively challenged is extremely damaging to these individuals. While these children and adults likely have neurological differences, their limited access to communication and education is much more disabling than anything else.”

Professional

“The underestimation of communication abilities has become more obvious with the advent of newer augmented communication techniques.”

Expert panel



A PARENT'S PERSPECTIVE

Tilly was 18 months old when diagnosed with Rett syndrome and 24 months when she started using an eye-gaze device. We always believed Tilly was intellectually aware, despite being told she was significantly delayed.

Augmentative and alternative communication (AAC) has given us a window into what she could achieve, from initially doing cause-and-effect games on an eye-gaze device and after a few minutes using it to ask for bubbles. Then asking to play with her friends at nursery as well as talking us through her day.

We always believed in Tilly; we modelled with symbols, demonstrating how to communicate with her, using both the eye-gaze device and low-tech communication book. I established a best “yes” and “no” using movements Tilly finds easy to make with her head.

It wasn't and it isn't easy, but consistency is key because Tilly understands and needs to feel valued, heard, and understood. We model and communicate, giving her options of how she wants to communicate so when she wants to talk, she can; when she wants to play games, she can; or when she wants to just have access to her voice, it is there and available.

Tilly knew how to use eye gaze immediately and asks for her food, asks to paint and draw, ignores you and puts it on rest, tells you it is hers, and tells you she is tired or uncomfortable. She finds symbols I don't even know exist and they make sense in the context. She has only just turned three!

Medical professionals said Tilly would never have met the criteria for an eye-gaze device and wouldn't have put her forward for funding, but we are pleased we did it anyway with the support and loan device from Rett UK. Now Tilly's medical team sees what Tilly is achieving, and it has helped them understand progress made in this area with people with Rett.

Thanks to Rett UK, without them I dread to think where we would be. Screaming has stopped, hope is restored, communication is key.

Collaborative and partnership working were essential to success. Tilly's speech therapist came to the Rett UK communication course with me; she said seeing what Tilly could achieve was the reason she became a therapist in the first place.

Her paediatrician has been hugely supportive and is always open to hearing about what we have learned.

Self-belief, belief in your child, the confidence to positively challenge professional opinion, working in partnership with professionals, modelling, modelling, modelling, and talking to Tilly in a variety of ways using a variety of AAC methods are what has worked for us. Whether that is eye gaze, intensive interaction, low-tech communication, or partner-assisted scanning, but always having her voice available to her.

Consistency is key, it isn't easy, and nothing worth having ever is. But communication and being heard are a basic human right, Tilly just accesses it in a different way than us, but she is proving to all of us just how effective it is.

